1999 SENATE HUMAN SERVICES

SB 2374

1999 SENATE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB2374

Senate Human Services Committee

❑ Conference Committee

Hearing Date JANUARY 25, 1999

Tape Number	Side A	Side B	Meter #
1		X	4,998
2	X		
2/1/99 2		X	4,985
Committee Clerk Signature	Carol Kolodejchuk		

Minutes:

The hearing opened on SB2374

SENATOR WAYNE STENEHJEM, sponsor, introduced bill and supports the bill. This bill will ask insurance companies to pay for help with the special dietary needs. The Senate passed the bill in great form, but the House put a sunset clause on it. We are asking that the sunset clause be removed and we pass the bill as it left the Senate two years ago.

REPRESENTATIVE KLINISKY, sponsor, stated that claims of $2000 were submitted to insurance companies; however, due to rewrites on policies we anticipate more claims in the next two years and following years to come. In the past biennium there was one child born with PKU so it is a very rare disease. There are some amendments proposed and I am concerned about the possibility that only two families would benefit from these amendments. SENATOR DEMERS: Why was the sunset clause put on two years ago? REP KLINISKY: It was because they thought

they needed some time and see what happened. Just to make sure the mandate didn't break the insurance companies backs; which with $2000 we didn't do.

REPRESENTATIVE POOLMAN, sponsor, agrees they will follow the Senate's infinite wisdom and take the sunset clause off. Has not seen the amendments, so we would request a look at the amendments before you act on them.

SENATOR LEE, also a sponsor, agreed that wonderful things will be done with proper food, and supports the bill.

SENATOR THANE, also a sponsor, support the bill two years ago and support it now.

SENATOR TIM FLAKOLL, sponsor, also supports bill. This past year a baby formula cost the family $7500 for the child. I can also be a reference as I am a biochemist.

MIKE MULLEN, Department of Health, written testimony, has presented an amendment. The department has provided formula to any child in need, but not food. SENATOR THANE asked how it was determined that a child has this disease. MR. MULLEN answered that blood tests taken in the hospital and within 3-6 months were placed on the special formula. SENATOR THANE asked is 185% of poverty level of WICK requirements was adequate. MR. MULLEN stated that there are several percentages to consider, but this will cover most of families and insurance will cover the rest. SENATOR DEMERS: Why did you choose WICK, because it floats with the number of applicants. Why not just pick a criteria and use it? Mr. MULLEN: I picked that because I thought it would be a well established standard. SENATOR DEMERS: Why not CHIPS? MR. MULLEN: We considered that option; maternal and child health staff recommended, also Health Office, on setting the higher standard of WICK level. It is a policy judgment on where it is to be set.

BRENDA RAKOCZY, mother of toddler with PKU, supports bill (written testimony). We have insurance coverage for lung cancer due to cigarette smoking, coverage for sexually transmitted diseases due to promiscuous behavior, and insurance coverage for drug and alcohol treatment centers. These are lifestyle choices. Individuals born with inherited metabolic disorders such as PKU, had no hand in their fate. They are innocent victims.

RENEE CHRISTIANS, mother of two children with PKU, supports bill with written testimony. Must keep on vegetarian diet to preserve mental health.

ANNE CHRISTIANS, College student with PKU, supports bill.

DAVE ZENTNER, Department of Human Services, determined that the department provides formula. This bill would mandate Medicaid to pay for the food and formula. SENATOR DEMERS asked about Dept of Human Services provides formula and Dept of Health formula. MR. ZENTNER: No, Dept of Human Services provides formula and Dep. of Health provides both formula and food.

DAN ULMER, BC/BS insurance rep, stated two people put claims in for food for $2035 and $376.29. We are not sure it was that effect. We anticipated more. This is something you are sure you cover. It should be done on a Health Dept. level. SENATOR DEMERS: Do you cover PERS? Yes, but I think it ends, so I don't think they are covering it right now. SENATOR DEMERS: Do you agree with the amendments? Mr. ULMER stated that he hadn't seen the amendments. Ms. RAKOCZY volunteered that many families wanted to put claims in but were told that due to the rewrite date they could not. Mr ULMER: Most of bills cycled after this date so that would be completely wrong, except for PERS.

DAVID AAKRE, parent of four children, three diagnosed with PKU. supports this bill with written testimony.

STACY BLACKSTEAD, PKU diagnosed, supports bill with written testimony.

MYRNA BLACKSTEAD, mother of PKU diagnosed child, supports bill with written testimony.

DANIELLE STRUXNESS, 17 year old diagnosed with PKU, supports bill with written testimony.

No opposition on the bill.

The hearing was closed; the committee adjourned until 9:00AM Tuesday, January 26, 1999.

The committee was reconvened on 2/1/99. Primary responsibility needs to be defined.

SENATOR LEE: This is responsibility of Health Department budget. SENATOR DEMERS: Health Department supplied formula; not food. SENATOR LEE and SENATOR DEMERS are to be a sub committee to report back ASAP.

MIKE MULLEN, Dept of Health, offered amendments to say that the insurance mandate does not require coverage for the low protein modified food products or medical food for the individual that has PKU or some metabolic disease. Those benefits are available under a program of the Health Dept such as the WIC, BLOC grant program or Medicaid. (attached testimony) SENATOR KILZER asked about the fiscal note. MR. MULLEN: An estimate is approximately $30,000, based on 15-17 children and $2000 average. SENATOR KILZER said it may be a little more. SENATOR LEE asked who does food and who does formula? MR. MULLEN: The Dept of Health would provide the formula and for those medically needed would provide the supplemental food. For children up to age 22 and for pregnant women, the Dept of Human Services. SENATOR DEMERS: Some of the people may be over income. MR.

MULLEN: That is when insurance would kick in and also for boys still going to school.

SENATOR DEMERS asked how many of the 18 total would be on insurance. Mr MULLEN: There is a list of persons on insurance. SENATOR FISCHER asked about the tracking of PKU. MR. MULLEN answered that there was a roster or register of all families. This is a confidential list, however. SENATOR THANE asked if there was a match from state funds. MR. MULLEN: They are all Federal funds. SENATOR LEE thanked the Health Department on their work in resolving this problem.

SENATOR DEMERS moved the amendments. SENATOR LEE seconded the motion. Roll call vote carried 6-0-0. SENATOR DEMERS moved a DO PASS AS AMENDED. SENATOR LEE seconded it. Roll call vote carried 6-0-0. SENATOR LEE will carry the bill.

FISCAL NOTE

(Return original and 14 copies)

Bill / Resolution No.: SB 2374 Amendment to:

Requested by Legislative Council Date of Request: 01/21/99

1. Please estimate the fiscal impact (in dollar amounts) of the above measure for state general or special funds, counties, cities, and school districts.

Narrative: This bill requires the Department to provide coverage for foods and food products for inherited metabolic diseases. The Department currently pays for these services and the costs are included in the 1999-2001 budget request contained in SB 2012. The actual cost of paying for these services is unknown as the Department does not track nutritional supplements by diagnostic code. Therefore, we can not separate payment for those required under this bill from those paid for other more common disease states.

2. State fiscal effect in dollar amounts:

	1997-1999 Biennium		1999-2001 Biennium		2001-2003 Biennium	
	General Fund	Special Funds	General Fund	Special Funds	General Fund	Special Funds
Revenues:						
Expenditures:	-0-		Unknown		Unknown	

3. What, if any, is the effect of this measure on the appropriation for your agency or department:

a. For rest of 1997-99 biennium: -0-

b. For the 1999-01 biennium: Unknown

c. For the 2001-03 biennium: Unknown

4. County, City, and School District fiscal effect in dollar amounts:

1997-1999 Biennium			1999-2001 Biennium			2001-2003 Biennium		
Counties	Cities	School Districts	Counties	Cities	School Districts	Counties	Cities	School Districts
-0-								

If additional space is needed, attach a supplemental sheet.

Signed Brenda M. Weisz

Typed Name Brenda M. Weisz

Date Prepared: January 22, 1999

Department Human Services

Phone No. 328-2397

Date: 2/10/99
Roll Call Vote #: 1

1999 SENATE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. 2374

Senate HUMAN SERVICES COMMITTEE Committee

☐ Subcommittee on _____
or
☐ Conference Committee

Legislative Council Amendment Number 90624.0101 .0200

Action Taken Amendment

Motion Made By Sen DeMers Seconded By Sen Lee

Senators	Yes	No	Senators	Yes	No
Senator Thane	✓				
Senator Kilzer	✓				
Senator Fischer	✓				
Senator Lee	✓				
Senator DeMers	✓				
Senator Mutzenberger	✓				

Total 6 (yes) 0 (no)

Absent 0

Floor Assignment _____

If the vote is on an amendment, briefly indicate intent:

state dept amendment

Date: 2/10/99
Roll Call Vote #: 2

1999 SENATE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. 2314

Senate HUMAN SERVICES COMMITTEE Committee

☐ Subcommittee on _____
or
☐ Conference Committee

Legislative Council Amendment Number 90624.0101 .0200

Action Taken Amended Do Pass

Motion Made By Sen DeMers Seconded By Sen Lee

Senators	Yes	No	Senators	Yes	No
Senator Thane	✓				
Senator Kilzer	✓				
Senator Fischer	✓				
Senator Lee	✓				
Senator DeMers	✓				
Senator Mutzenberger	✓				

Total 6 (yes) 0 (no)

Absent 0

Floor Assignment Sen Lee

If the vote is on an amendment, briefly indicate intent:

REPORT OF STANDING COMMITTEE

SB 2374: Human Services Committee (Sen. Thane, Chairman) recommends **AMENDMENTS AS FOLLOWS** and when so amended, recommends **DO PASS** (6 YEAS, 0 NAYS, 0 ABSENT AND NOT VOTING). SB 2374 was placed on the Sixth order on the calendar.

Page 1, line 1, remove "a new section to chapter 50-24.1 and"

Page 1, line 2, remove "medical assistance and"

Page 2, after line 12, insert:

"4. This section does not require medical benefits coverage for low protein modified food products or medical food for an individual to the extent those benefits are available to that individual under a department of health program or under a department of human services program."

Page 2, remove lines 13 through 18

Renumber accordingly

1999 HOUSE HUMAN SERVICES

SB 2374

1999 HOUSE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB 2374

House Human Services Committee

❑ Conference Committee

Hearing Date March 8, 1999

Tape Number	Side A	Side B	Meter #
1		x	38.6-End
2	x		0.0-44.2
3	x		10.1-18.3
Committee Clerk Signature			

Minutes:

Senator JUDY LEE, District 13, testified. (Testimony attached.)

Representative AMY KLINISKE, District 42 testified in support of the bill. Rep. AMY KLINISKE said that the bill will provide seamless coverage for the affected individuals in conjunction with Department of Health, Department of Human Resources and Medicare and removes the sunset clause.

DAVID ZENTNER, Director of Medical Services for the Department of Human Services appeared to provide information and offer an amendment to the bill. (Testimony attached.)

MURRAY SAGSVEEN, State Health Officer testified. (Testimony attached.)

ALAN KENIEN from Fargo testified in favor of the bill. (Testimony attached.)

BRENDA RAKOCZY testified in favor of the bill. (Testimony attached.)

NANCY OUDERKIRK testified in favor of the bill. (Testimony attached.)

ANNE CHRISTIANS from Fargo testified. (Testimony attached.)

DAVID AAKRE testified in favor of the bill. (Testimony attached.)

DANIELLE AAKRE testified. (Testimony attached.)

Dr. KATHY WOOD a physician from Grand Forks testified. (Testimony attached.) In response to questions from the committee Dr. WOOD provided additional information. Children with the disease must remain on the diet for life. At one time it was thought that after the age of six the special diet could be discontinued. It has since been learned that deterioration to the mental capabilities will begin at any age that the diet is discontinued. If the diet is restarted conduct and mental capacities can return. However, any brain damage is permanent.

STACY BLACKSTEAD testified. (Testimony attached.)

There was no OPPOSITION to SB2374.

Hearing closed on SB2374.

SB2374 reopened for committee action.

Rep. AMY KLINISKE moved to amend the bill by removing "or under a" from page 2, line 13 and removing "department of human services program" from page 2 line 14. Rep. CAROL NIEMEIER seconded. Motion PASSED, Voice vote: 15-0-0.

Rep. AMY KLINISKE moved "DO PASS AS AMENDED". Rep. CAROL NIEMEIER seconded. After some discussion on coverage, eligibility and rules for coverage the motion was voted on.

Motion PASSED on Roll Call vote #6: 10 Yes, 5 No, 0 Absent.

CARRIER: Rep. PAT GALVIN

Date: 3/8/99
Roll Call Vote #: 6

1999 HOUSE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. SB2374

House Human Services Committee

☐ Subcommittee on _____
or
☐ Conference Committee

Legislative Council Amendment Number ~~SB2374~~

Action Taken Do Pass as Amended

Motion Made By Rep Kliniske Seconded By Rep Niemeier

Representatives	Yes	No	Representatives	Yes	No
Clara Sue Price - Chairwoman	✓		Bruce A. Eckre	✓	
Robin Weisz - Vice Chairman		✓	Ralph Metcalf	✓	
William R. Devlin		✓	Carol A. Niemeier	✓	
Pat Galvin	✓		Wanda Rose	✓	
Dale L. Henegar	✓		Sally M. Sandvig	✓	
Roxanne Jensen	✓				
Amy N. Kliniske	✓				
Chet Pollert		✓			
Todd Porter		✓			
Blair Thoreson		✓			

Total **Yes** 10 No 5
Absent 0

Floor Assignment Rep Galvin

If the vote is on an amendment, briefly indicate intent:

REPORT OF STANDING COMMITTEE

SB 2374, as engrossed: Human Services Committee (Rep. Price, Chairman) recommends **AMENDMENTS AS FOLLOWS** and when so amended, recommends **DO PASS** (10 YEAS, 5 NAYS, 0 ABSENT AND NOT VOTING). Engrossed SB 2374 was placed on the Sixth order on the calendar.

Page 2, line 13, remove "or under a"

Page 2, line 14, remove "department of human services program"

Renumber accordingly

1999 TESTIMONY

SB 2374

MeritCare

MERITCARE HOSPITAL
720 4TH ST. N.
FARGO, ND 58122

MERITCARE MEDICAL GROUP
737 BROADWAY
FARGO, ND 58123

January 19, 1999

North Dakota State Assembly of the Government

I, Alan Kenien, strongly favor the passage of this legislation to help families who have a member with an inherited metabolic disease. I am a board certified pediatrician and pediatric endocrinologist and am the medical director of the State Metabolic Disease Clinic who sees most of the patients in the state of North Dakota with inherited metabolic diseases. These diseases affect the newborn and continue to influence the child throughout the child's lifetime. Metabolic diseases often cause mental retardation. Some of the metabolic diseases also cause life-threatening illnesses during the newborn period and during and after the newborn period. The most common inherited metabolic disease in North Dakota is PKU. There are approximately twenty individuals in North Dakota who have PKU who are followed at our clinic. In this disease, the body is unable to utilize an amino acid that we eat in any foods that contain proteins. As a consequence, the body's level of this amino acid (phenylalanine) accumulates to very high levels. The phenylalanine excess leads to mal-development of the brain, which subsequently leads to mental retardation, if the disease is not identified and treated by the age of three months. We are fortunate in the state of North Dakota as in the rest of the United States and other developed countries to have a screening program which detects PKU at birth. Once an infant is identified with this disease, the infant can be successfully treated with a special formula which the state has so generously provided in the past. Instead of being able to eat protein-containing foods such as milk, meat, eggs, and many other foods, this formula must be substituted. As the child grows the formula does not provide all of the nutrition that the child needs and other foods must be introduced into their diets. These are the "low protein modified products". These low protein modified products are extremely expensive and the temporary House bill that was passed two years ago helps to provide financial assistance for families to help pay for these low protein modified products. If the families are unable to afford these foods and give their children foods that other children eat, their phenylalanine levels will rise so high that it will affect their mental development and behavior. It has been shown that children who are taken off of their diets significantly lose IQ points. It has also been shown that these children develop behavior problems and learning problems such as attention deficit disorder.

I am strongly in favor of making this law permanent so that children with these rare diseases can be helped in order to give them an equal chance to develop and thrive and in this way, have an equal chance as normal children. If you need any further information about this condition or it's treatment, I will be happy to elaborate. I hope to be able to attend the hearing if my schedule and the weather permits.

Sincerely,

Alan Kenien

Alan G. Kenien, M.D.

krb

Children's Hospital
300 Longwood Avenue, IC-106
Boston, MA 02115
617-355-6346 (tel)
617-730-0461 (fax)
levy_h@a1.tch.harvard.edu (e-mail)

Harvey L. Levy, M.D.
Senior Associate in Medicine/Genetics

Children's Hospital

January 11, 1999

Dear Members of the Human Services Committee:

It is my understanding that Senate Bill 2194 that requires health insurers to cover the low protein modified food and metabolic formulas for individuals with phenylketonuria (PKU) and maple syrup urine disease (MSUD) has a two-year sunset clause. This is an extremely important bill and I would strongly advocate removing that clause. A child with PKU or MSUD has a very serious disease for which the formula and low protein foods are as critical treatment for them as is insulin for a child with diabetes or steroids for a child with another chronic disease.

When a child with PKU is not properly treated, severe mental retardation results. Not only would this be an inexcusable tragedy for the child or family but it would result in great expense to them and to the health insurer for coverage of the needs of a child with mental retardation. In cases of MSUD, a child who is not properly treated will not only suffer severe mental retardation but will also require numerous hospitalizations and intensive care for serious metabolic episodes, all of which would be tragic and, again, extremely expensive. In addition, a child with MSUD who is not treated could die. In short, quite aside from the humanitarian needs, proper treatment for these diseases is far less expensive than the consequences of inadequate treatment.

Many states have now passed laws such as the one you passed in North Dakota which require coverage by health insurers of these treatments. To my knowledge, none other than the North Dakota bill has a sunset clause. I urge you to remove that clause on behalf of the children and their families who so deserve this support for treatment of their serious diseases.

Please feel free to call me at 617-355-6346 or fax me at 617-355-3461 if there are any questions.

Sincerely,

Harvey L. Levy, M.D.

UNIVERSITY OF UND NORTH DAKOTA

SCHOOL OF MEDICINE & HEALTH SCIENCES
DEPARTMENT OF PEDIATRICS
DIVISION OF MEDICAL GENETICS
501 NORTH COLUMBIA ROAD
P.O. BOX 9037
GRAND FORKS, NORTH DAKOTA 58202-9037
PHONE (701) 777-4277
FAX (701) 777-6124

January 11, 1999

To Whom It May Concern:

It has come to my attention that Senate Bill 2194 had a sunset clause that necessitates a reexamination and extension of this particular bill or eliminating the sunset clause from the previous bill. I am in support of removing the sunset clause from Senate Bill 2194 as proposed in Senate Bill 2314 that requires health insurance companies to provide up to $3000 coverage per person per year for individuals with metabolic disorders. This is in particular those metabolic disorders for which a specific dietary treatment is available. In the past this has included phenylketonuria and maple syrup urine disease. However, it would be important to not limit the coverage so that as metabolic disorder treatments improve to the extent that they have with phenylketonuria and maple syrup urine disease, that these diseases would also be covered. However, at this particular time it is important to continue the policy set forward in the previous Senate Bill 2194.

The individuals that are presently or will be in the future receiving medical dietary treatment for particularly PKU and maple syrup urine disease, develop intellectually and become productive members of society. Without the diet, these individuals would be intellectually impaired and would not be able to be productive tax paying members of society. Individuals on the diet have normal or close to normal intelligence and are able to function quite well within the regular school systems and to go on to college and future educational levels. Without the diet this is impossible. Also, the diet prevents certain psychiatric disorders that can occur in untreated individuals, particularly with PKU. This particular diet is life long, since women who have phenylketonuria that become pregnant and do not maintain their diet have, 100% chance of having mentally retarded children, who may or may not have phenylketonuria themselves. In other words, the mother who does not maintain her treatment for phenylketonuria during pregnancy can have high levels of phenylalanine that can adversely affect any fetus that she is carrying. Therefore, in order to provide the best outcome for the pregnancies in these particular mothers, it is imperative that they stay on the diet for life. Male individuals with metabolic disorders for whom a diet is prescribed perform better in society if they stay on the diet life long. The particular dietary products are a supplement to a modified diet that these individuals need to take in order to prevent the elevation of the abnormal products that can result from eating a full regular diet. The medical nutritional diet allows them to have a well balanced diet from the standpoint of protein and carbohydrates that would not otherwise be available. The diet allows them to develop normally.

Therefore, I strongly urge support for the removal of the sunset clause of the previous Senate Bill 219[4] as expressed in the present Senate Bill 237[4]

If there are additional questions or comments, please do not hesitate to contact me.

Sincerely yours,

John T. Martsolf, MD
Division of Medical Genetics

JTM/jd

Altru
Health System

January 14, 1999

Dear Senators:

I am writing in support of the "Food and food products for inherited metabolic diseases" bill currently being considered in the Fifty-fifth Legislative Assembly. As a psychologist who treats children with metabolic disorders, I am asking you to lend your support to this bill. There are people with metabolic disorders in North Dakota who do not have the proper diet available to them because of the extremely high cost of these foods. This has the immediately apparent effect of higher health care costs for these patients. There are also long term effects related to resulting mental retardation and learning disabilities that ultimately costs our state.

I urge you to carefully consider the benefits of supporting this legislation. This bill will help make it possible for all persons to develop up to their potential.

Sincerely,

Eileen Searcy, Ph.D.
Clinical Psychologist

Altru Rehabilitation Center
Altru Health Institute ■ 1300 South Columbia Road ■ P.O. Box 6002 ■ Grand Forks, ND 58206-6002 ■ 701-780-2311

The Arc, Upper Valley

P.O. Box 12420
2500 DeMers Ave.
(701) 772-6191 Office Grand Forks, ND 58208-2420 Fax (701) 772-2195

January 19, 1999

Members of the Senate and House Human Service Committee:

The Arc, Upper Valley is an organization committed to improving the quality of life for children and adults with mental retardation and related disabilities and their families through advocacy, education and family support services. We would like to voice our support for proposed Senate Bill #2374. This proposed legislation would extend the current health insurance coverage of metabolic formula and low protein modified foods to families who have children needing this type of medical treatment.

This proposed legislation would also ensure that insurance companies continue to provide coverage for metabolic formula and low protein modified foods for PKU(Phenylketonuria) and MSUD(Maple Syrup Urine Disorder) patients by removing the sunset clause contained in Senate Bill 2194 which became law in 1996. The continuing coverage of metabolic formula and low protein modified foods under insurance policies supports mental retardation prevention efforts. Mental retardation and other neurological problems can develop if proper nutrition therapy is not maintained. We urge you to support this important Bill.

Sincerely,

Peter Johnson
Chair, Governmental Affairs Committee
The Arc, Upper Valley

Virginia Esslinger
President
The Arc, Upper Valley

IN SUPPORT OF SENATE BILL 2374
INDEX OF TESTIMONY

MEDICAL TESTIMONY

1. Doctor Eric Lunn Grand Forks, ND	American Academy of Pediatricians
2. Doctor Alan Kenien Fargo, ND	Pediatric Endocrinologist – Meritcare Hosp
3. Doctor Harvey Levy Boston, MA	Senior Associate in Medicine/Genetics Children's Hospital
4. Doctor John Martsolf Grand Forks, ND	UND School of Medicine – Dept. of Pediatrics Division of Medical Genetics
5. Doctor Eileen Searcy, PH.D Grand Forks, ND	Altru Health Systems – Clinical Psychologist

NATIONAL ORGANIZATION

The ARC, Upper Valley – Peter Johnson Virginia Esslinger - Grand Forks, ND	Chair, Governmental Affairs CMTE President, ARC

PARENTS OF CHILDREN WITH PKU AND INDIVIDUALS WITH PKU

1. Brenda Rakoczy Reynolds, ND	Mother of toddler with PKU
2. Ann Christian Fargo, ND	College student with PKU – NDSU
3. Renee Christian Fargo, ND	Mother of two children with PKU
4. David & Aakre & daughter Danielle – 12 Kindred, ND	Father of 4 children, 3 with PKU
5. Mrs. Myrna Blackstead Bismarck, ND	Mother of PKU Adult
6. Stacey Blackstead Bismarck	PKU Adult
7. Sheila Struxness & daughter Danielle Hazen, ND	Mother & teenage daughter with PKU

8. **Nancy Ouderkirk**
   Fargo, ND
   Mother of a toddler with PKU

9. **Gayle Heskin**
   Minot, ND
   Mother of teenager with PKU

10. **Dale & Kris Wangler**
    Rugby, ND
    Parents of the Youngest Baby with PKU

11. **Marshall & Tammy Anderson**
    Manvel, ND
    Parents of toddler with PKU

12. **Kim & Brenda Ankenbauer**
    Washburn, ND
    Parents of a teenager with PKU

13. **Jerry Wollenberg**
    Hamiliton, ND
    Mother of a preschooler with PKU

14. **Keith & Linda Ulmer**
    Bismarck, ND
    Parents of a preteen with PKU

MAPLE SYRUP DISEASE

Amy & Jeff Johnson
Fargo
Parents of a 7 year old boy with MSUD

My name is Anne Christians. I am 22 years old. I am a senior at NDSU. I am blond and have blue eyes. I am also a phenylketonuric. This means I have PKU. This is not so much what I have as who I am. I have been on a restricted diet my entire life as I will be forever. I am lacking an enzyme in my liver that processes a certain protein. Therefore, I am a strict vegetarian. I eat no meat, dairy products, eggs, or fish. I can't even drink diet soda. I drink a special formula to provide my body with all the nutrients that I can't get through my diet. This formula is what has guaranteed me a "normal life". Without it I would have been mentally retarded. I feel very fortunate that I was born after PKU was diagnosed and a treatment had been developed. However, this treatment does not end. For me to maintain and continue my life as a successful student and productive member of society I need this formula. Without it my thought processes would become unclear and I would become lethargic and unmotivated. More severe consequences could be neurological damage and psychiatric problems. As a woman, this is an especially important issue to me, as I will also need this formula to ensure the health of my unborn children.

At the present time I am completing a one year internship in Medical Technology at Trinity Medical Center in Minot. After this summer I will be looking for a job. Please don't force me to have to leave the State of North Dakota. If the insurance coverage is allowed expire it would be difficult for me to afford the cost of the formula and to also start repaying my college loans. This a genetic disorder that I did not ask for, nor could I control being born with it. Please do not penalize me for this.

There are so many other things that I could say. I feel very lucky when I think about my life and all the wonderful things I've been able to see and do compared to how my life would have been without this diet, my formula and the people who care.

On behalf of myself, my brother, and all the other PKU children in the state I urge you to remove the sunset clause from Senate Bill No. 2194.

My name is Renee Christians. I'm a native North Dakotan and live in Fargo. I am the mother of two PKU children. My children, Anne, age 22, and Daniel, age 20, are the oldest PKU children in the state who have been on this restricted diet since birth. Their diet is based on their formula and a strict vegetarian program, excluding almost all protein. Foods you and your families take for granted my children have never tasted. Anne and Daniel have never had a hamburger, a Snickers candy bar or even a glass of milk. However, this is a small price to pay for mental health. It would be impossible to tell you about 20 years in the space of a few minutes, but I can testify to the fact that because my children have been on this diet all their lives, they are intelligent, healthy, well-adjusted individuals, on their way to becoming productive citizens and taxpayers. Without this formula and diet my children would be severely mentally retarded. They would have had no future. As it is they are both excellent students. Anne is a senior at N.D.S.U., majoring in Medical Technology. Daniel, a junior at U.N.D., is a Business major. They are the same as any other children. They both have dreams, goals, and ambitions.

I would like to thank the state of North Dakota, and in particular the Maternal and Child Health Department, for providing a formula for my children that has allowed them to be able to grow and develop, both mentally and physically, and to reach their full potential. The expense of this formula would be a hardship for many families, my family included. When Anne was born there was no way that we could have afforded the cost of this formula. Having two children on this formula costs about $10,000 a year, and that is just for the formula. I thank God the state of North Dakota has paid for it. However, now that Anne is 22 and Daniel will be 21 this summer, the state will no longer provide this formula. It has become imperative that insurance companies be

held responsible for the cost. I am asking you now to remove the sunset clause and to make this a permanent law. PKU formulas are by prescription only, and as such should be covered by insurance. The insurance companies may argue that these formulas are a food and not a medicine. I strongly disagree. As a "food", I don't know of any family that pays $10,000 a year for milk . Without this "food" my children would be mentally retarded. I can't think of one other "food", that if withheld, would have those results. They need this formula to be able to achieve and live a normal healthy life. If the sunset clause is removed and this bill stands, our family would still be responsible for $4000 a year for formula. If this bill is allowed to expire, we would be responsible for the full $10,000. If that is the case, I would urge my children to leave the state of North Dakota and seek employment elsewhere, in a state that does cover PKU.

Although their brains are developed, my children need this formula and to be on this diet, now as much as ever. It has been documented in adults that without this diet there can be neurological problems even to the point of becoming psychotic. Their thought processes slow down and become fuzzy and unclear. They can become depressed and lethargic. Physically, they could have seizures, headaches and sleep disorders. They have been known to lack judgment and reasoning, leading to crimes and jail sentences. Not only as college students, but for the rest of their lives, my children need to be sharp, clear and focused.

Maternal PKU is also a very real concern as my daughter is now an adult. If Anne was not on the diet, she would have elevated levels of Phenylalanine in her blood. As a pregnant woman, Anne's body would supply the baby with its nourishment. Because the baby would feed through Anne's bloodstream, it would be born retarded even though it would not have PKU. She will need to be on this diet, with the formula,

throughout her child bearing years. On the diet she will be able to have normal healthy babies. Without it, she will give birth to retarded children. This is a fact.

The long term ramifications of not having this formula paid for by insurance companies are immense. Potentially, we could have retarded children who would be a responsibility of the state. It is mandated that all children receive an education to meet their needs. The expense for educating these children would be enormous. And then what, children in institutions. Can you imagine what that would cost? My husband and I love our children dearly, and as parents, we want what is best for them and would do anything to provide for their needs. But sometimes the expenses are so great that many families would not be able to afford the cost of this diet. The states return on their investment in the health of PKU children is profound. Intelligence or retardation. There is no choice.

When the answer is so clear, why would we take the chance of not providing these children with the means of obtaining this formula. Please, I urge you to remove the sunset clause from Senate Bill No. 2194 so that insurance companies to be held responsible.

Chairman, members of the Human Service Committee, my name is Brenda Rakoczy. Thank you for allowing me the opportunity to testify today on behalf of my family and the North Dakota ARC, in support of Senate Bill 2374. I am the mother of Sydney Marie who is three years old and has PKU. Sydney was diagnosed with PKU trough **mandated** newborn screening. She has been tested by a clinical psychologist and an educational specialist as quite advanced for her age. She is reaching her full physical and mental potential because of a well-maintained, medically regulated low protein diet. She is one of the many PKU success stories you see in this room.

As you are aware undected and untreated PKU will result in severe mental retardation in infancy. If children with PKU are not able to maintain a strict dietary regimen for life they become victims of slow, but quite significant intellectual and neurological deterioration over a period of time. This terrible development can be prevented by following a very strict low protein, vegetarian diet which includes specially prepared prescription metabolic formula and low protein modified foods to provide the bulk nutrients lacking in their highly restricted diet. Sydney must restrict all meat, dairy, poultry, seafood, legumes, nuts, and most products that contain flour. A typical diet includes small portions of preciously weighed fruits and vegetables low in protein.

Sydney is only allowed 250 mg of phenylalnine (phe) per day. Let me put that into perspective. If I allowed Sydney this one piece of bread which is 110 mg of phe and ½ cup of honeynut cheerios (106) – with out milk of course, it would use up 216 of her 250 mg of protein allowed per day. That would only leave her with 44 mg of protein for her lunch, supper and snack. Without the low protein modified foods she basically would go hungry. Even fruits and vegetables are limited in their amount. 1 ear of corn on the cob is 256 mg of phe, ¼ cup of peas are 75, 1 medium banana is 43 mg of phe, and 1 small fry at Burger King 140.

We think of everything that goes in her mouth as a medicine, and it is a constant challenge to meet her hunger, calorie and nutritional needs without adding harmful phenylalnine. Families must keep carefully maintain diet menus and daily logs of all the medical formula and foods that goes into their children mouth. See ACTH 1. An often-ignored fact about the PKU diet is that the child's allowance for phenylalanine may not increase, as they grow older. On the other hand, Sydney's appetite will surely increase.

The heart of the problem now encountered by a family whose child is diagnosed with PKU is lack of insurance to cover the high cost of these prescription formula and low protein modified foods, they remain cost prohibited to most. The low protein modified foods broaden the variety of foods an individual may use to maintain dietary control. This is particularly important as authorities recommend the diet should be lifelong. Unfortunately these products are quite costly and are only manufactured by four companies in the United States. One box of loprofin mix to make a loaf of bread which is half the size of our normal bread is $6.55, a 17.6 oz box of low pro rice is $7.75, small box of cracker $6.83, 17 oz box of elbow macaroni, 7.75, 3 ounce imitation Jell-O $1.35 and a 9 ounce box of imitation lo protein chocolate $9.85. This does not include the shipping and handling. In summary, these special low protein foods make a very difficult dietary regimen tolerable and improve the medical well being of many of our children.

Than there is the cost of the prescription metabolic formula for Sydney which is $158.00 for 6 cans, she goes through approximately 12 cans per month at $316.00 and than the metabolic flavoring to disguise the terrible taste of the formula is $46.00 for 6 cans. With upcoming cuts from the State Health Department on distribution of prescription formula to families, and with out the assistance of our insurance companies, there is no way possible our family can afford $433.00 per month for Sydney prescription medical food. Please understand this does not include our monthly health insurance

payment or other cost associated with raising a child. The cost continues to grow as her body changes and requires more. Yearly the cost of the products continue to rise. Without this assistance her treatment may be delayed or inadequate. A public policy that would allow a PKU or MSUD individual to a state of irreversible and variable mental damage would have to be perceived as morally unacceptable.

At one time it was common practice to take a child off the diet after their brain was fully developed. Clinical studies have proven that this was a mistake and individuals who come off the diet during anytime of their life stand a high chance of developing continued neurological deterioration, loss of intellectual development and mental problems. **Abandoning their diet will put them on a dangerous track.** Aside from the humanitarian needs, proper treatment for these disease is far less expensive than the consequences of inadequate treatment. In North Dakota we spend $339 dollars per day at Developmental Center for one individual, the cost for one client in a group homes runs approximately $182.00 per day and than there is the cost of special education which continues to rise on a upward pendulum.

With Sydney we also have the maternal PKU issue when she decides to marry and have children. Women who are not on a highly restricted diet for at least a year prior to conceiving have a very high chance of harming their unborn baby. Untreated maternal PKU causes defects in offspring, including severe mental retardation, heart defects, complications of pregnancy, fetal loss and induced abortions.

PKU screening has become compulsory since 1963 in the United States. Is our state not obligated to assure that the prescribed medical resources necessary for effective treatment are made available. As Doctor Lance Wyble stated in his testimony to the Florida Senate on behalf of mandated insurance coverage for inherited metabolic disorders such as PKU. **"It is important to note that any health coverage which in theory has been purchased for the express purpose of treatments of medical conditions owns certainly no less a responsibility than the State institution which mandates diagnoses."**

North Dakota families are doing their best with the physical, psychological, dietary and financial pressures of this disease, but need your help. Passage of this bill would alleviate this problem with an obvious workable effect on the insurance industry due to the extremely low incidence rate. Additionally, most plans provide for the 80 – 20 split. On top of the monthly health insurance cost, we pay the shipping and handling fees. We have only had one child born with PKU in our state since passage of Senate Bill 2194. We respectfully request that the two-year sunset clause be permanently removed. If this bill is passed – families could than concentrate on the effective medical therapy for their children.

As a parent of a loving, well adjusted, active toddler with PKU, we want to allow her the opportunity to grow up healthy, happy and with the ability to set and accomplish goals and dreams. Regardless if it's flipping hamburger at Burger King, going to college, or becoming a congressional leader for the State of North Dakota, we just want to give her the basic fundamental rights and abilities to decide.

In conclusion, I leave you with these thoughts; in America we have insurance coverage for lung cancer contributed by cigarette smoking, coverage for sexually transmitted diseases due to promiscuous behavior, and insurance coverage for drug and alcohol treatment centers. Please know I'm not passing judgment on these individuals. However, **they are all lifestyle choices. Individuals born with inherited metabolic disorders such, as PKU had no hand in their fate they are innocent victims.** Because infinite needs have encountered finite resources, we must ~~no~~ Now plan our spending wisely.

Will we choice life style choices over a diabetics right to insulin, a women's right for breast cancer treatment or a child's right to be kept free from the devastating affects of severe mental retardation and continued neurological defects? Please I ask you to help keep these children healthy. Allow them to become tax providers for our state, not tax burdens. **Please vote yes on Senate Bill 2374.** Thank-you.

KEEPING TRACK OF MILLIGRAMS OF PHENYLALANINE

Diet Prescription: 225 mg phe Note: Method 1 and Method 2 are described on pp. 21-22.

Food You Want to Serve	Amount You Want to Serve	Food List Portion Size	What You Need to Do to Figure Phe	Total Phe (mg)
Breakfast				
Rice Krispies	1/2 cup (14 gm)	1/2 cup (14 gm)	*First, weigh 14 gm or measure 1/2 cup. Then, use book value.*	45
Orange Juice	8 fl. oz.	4 fl. oz.	*First, measure 8 fl. oz. Then, use Method 2: 8 ÷ 4 = 2 and 2 x 15 mg phe =30.*	30
			Sub-Total Breakfast	75
Lunch				
D. Specialties Low Pro. Bread	2 slices	1 slice	*Use Method 2: 2 ÷ 1 = 2 and 2 x 3 mg phe = 6.*	6
Steve's Peanut Butter (Low Protein) NO REAL PEANUT BUTTER ALLOWED	2 Tabl.	1 Tabl. = 3 mg phe (recipe from Low Pro. Bread Mach. Baking*)	*Use Method 2: 2 ÷ 1 = 2 and 2 x 3 mg phe = 6.*	6
Fresh Peach	1 med.	1 med (172 gm)	*First, weigh the peach. Say it weighs 135 gm. Then, use Method 1: .22 mg phe /gm food x 135 = 30.*	30
Mother's Circus Animals	2 cookies	1 cookie	*Use Method 2: 2÷ 1 = 2 and 2x 6 mg phe = 18.*	18
			Sub-Total Lunch	60
			Running Total (75 + 60):	135
Dinner				
Aproten Rigatini (Low Protein)	2/3 cup	2/3 cup (62 gm), dry	*Weigh 62 gm, or measure 2/3 cup.*	12
Hunt's Classic Spaghetti Sauce	1/3 cup	1/2 cup (125 gm)	*First, measure 1/3 cup. Then, use Method 2: 1/3 cup = .33 and 1/2 cup = .5; then .33 + .5 = .66 and .66 x 54 mg phe = 36.*	36
Carrot sticks	3 sticks	1 whole (90 gm) carrot	*First, weigh carrot sticks. Say they weigh 25 gm. Then, use Method 1: .32 x 25 gm = 8.*	8
Lettuce	1/2 cup torn pieces	1/2 cup (29 gm) torn pieces	*First, weigh 29 gm or measure 1/2 cup. Then, use book value.*	12
Italian Dressing	1 Tabl.	1 Tabl.	*Use book value.*	"Free"
Tiny Goldfish Crackers, Orig.	10 crackers	10 crackers	*Use book value.*	18
			Sub-Total Dinner	86
			Running Total (135 + 86):	221
Snack				
Sunkist Fun Fruits	1 pkg. (25 gm)	1 pkg. (25 gm)	*Use book value.*	3
			The Grand Total:	224

* See p. 29 for ordering book.

from Low Protein Food List for PKU by Virginia Schuett

Testimony
on
SB 2374, PKU Insurance Benefits
before the
Senate Human Services Committee
by
Michael J. Mullen, Department of Health

January 25, 1999

Good morning, Mr. Chairman and members of the Committee. My name is Michael J. Mullen, Senior Advisor for Health Policy with the Department of Health. I am pleased to present testimony regarding the Department's PKU screening and special formula food program.

Phenylketonuria (PKU) and Maple Syrup Urine Disease (MSUD) are two uncommon inherited metabolic diseases. The North Dakota Department of Health has conducted a metabolic screening program for PKU and provided special formulas to PKU children since the late 1960s. Children with PKU, if detected at an early stage, may grow to have normal intelligence if a moderately expensive dietary treatment is provided.

Phenylketonuria occurs in 1/10,000 births. Maple Syrup Urine Disease occurs in 1/200,000 births. North Dakota has approximately 8,500 births a year. Thus, on average, one child with PKU is born in North Dakota every 14 months. Formula costs for children with PKU who stay continuously on the formula are approximately $3,800 per year. Low protein modified food costs range from $1,500 to $2,500 per year according to information from other states. Formula costs for MSUD are $7,000 per year. The Department of Health currently provides special formulas for 17 children (under the age of 20) with PKU and one child with MSUD.

Approximately $100,000 of Maternal and Child Health [MCH] block grant funds will be used this biennium to purchase the needed formulas.

The Department of Health also collaborates with the Department of Human Services, Children's Special Health Services (CSHS) Division, formerly Crippled Children's Services, in conducting metabolic specialty clinics in North Dakota. These clinics provide multispecialty services funded with CSHS monies.

While special formula and low protein modified food products were previously recommended for children with PKU through preschool and the early elementary years when commencement of a regular diet would begin, some experts now recommend life-long treatment of PKU with these products. Under current law, the Department is authorized to use MCH block grant funds to provide the benefits only to PKU children up to age 22, as well as pregnant PKU women, and then (in either case) only if they are financially unable to pay for these special foods.

Mr. Chairman, as I previously mentioned, the State of North Dakota has for three decades required that children be screened for PKU. Experience has shown that nutritional therapy can effectively prevent severe mental retardation in children with PKU. The Department of Health has provided formulas and special foods that have helped many families and children over the years.

There is one amendment the Department would like to recommend. It appears that the Department has never issued any rules to objectively define individuals who are eligible the PKU formula benefit. Therefore, we are proposing an amendment to section 25-23-03(2) to define an individual eligible for PKU formula benefits as an individual who meets the income eligibility requirements for the special supplemental nutrition program for women, infants, and children

("WIC"), to the extent that medical assistance [Medicaid] or the Children's Health Insurance Plan [CHIP] do not cover these benefits. [A copy of the amendment is attached.]

Mr. Chairman, that concludes my formal testimony. I will be happy to answer any questions that you or other members of the Committee may have about the Department's PKU program.

# # #

PROPOSED AMENDMENT TO SENATE BILL NO. 2374

On page 2, after line 25, insert:

"SECTION 4. AMENDMENT. Subsection (2) of section 25-17-03 of the North Dakota Century Code is amended and reenacted as follows:

Make arrangements for the necessary treatment ~~for~~ of a diagnosed ~~cases~~ case of any individual ~~where~~ if treatment is indicated and the ~~family is unable to pay the cost of such treatment~~ meets the income eligibility requirements for the special supplemental nutrition program for women, infants, and children ("WIC") [P.L. 105-216, 42 U.S.C. § 1786], to the extent that medical assistance [Medicaid] or the Children's Health Insurance Plan [CHIP] do not cover these benefits."

TESTIMONY: North Dakota Chapter of the American Academy of Pediatrics
Eric R. Lunn, MD, FAAP
Senate Bill # 2374
January 11, 1999

Mr. Chairman, members of the committee:

Thank you for the opportunity to testify today. I sincerely apologize for being unable to testify in person. Due to conflicts in my clinical practice I am unable to attend your committee hearing in person. I am truly sorry for this inconvenience. My name is Eric Lunn, I am a pediatrician practicing in Grand Forks. I appear before you on behalf of the members of the North Dakota Chapter of the American Academy of Pediatrics, which you know as an international organization who, not only provide health care for children but share a longstanding deep commitment to be the voices for innocent children throughout the world. I also appear on my own right as a concerned pediatrician to express continued support for a law that was passed in 1997 which provides reimbursement for medical foods for certain inborn errors of amino acid metabolism.

As you are aware this law was passed in 1997 to provide support for these unfortunate children. I would like to briefly discuss what these disease are and what the disease cause to the individuals who have them and their families. They are a group of rare inherited disease that untreated culminate in severe mental retardation or death. Most of these disorders are abnormalities in the metabolism of amino acids which are building blocks for proteins. Inborn errors of amino acid metabolism include phenylketonuria (PKU), maple syrup urine disease (MSUD), and several other less common disorders. Treatment might include restriction of one or more amino acids in the diet, restriction of total protein in the diet, or a supplementation of specific substances such as vitamins. Again, untreated diseases result in severe mental retardation or death.

PKU is one of the most common disorders of amino acid metabolism and occurs in approximately 1 out of every 12,000 births. PKU is a disease that affects the way a person processes food. A child with PKU cannot process an amino acid called phenylalanine which is a building blocker for proteins. As a result, phenylalanine builds up in the blood stream and causes brain damage and mental retardation.

Children born with PKU appear normal for the first few months of life, however, untreated they begin to lose interested in their surrounds and by 3 to 5 months of age and by the time they are a year old are mentally retarded. Fortunately, all states now routinely screen for PKU. If a baby is found to have PKU and placed on a special diet low in phenylalanine before the fourth week of life, mental retardation can be prevented.

Once the diagnosis is confirmed, treatment for inborn errors of amino acid metabolism (including PKU and MSUD) must be carefully monitored by a physician with expertise in metabolic disease. Early in life, these children require specialized formula in which the protein has been modified. As they grow older, they are unable to eat normal protein containing foods such as milk, meat, eggs, and other foods. Special medical foods, including low protein and modified food products, are indispensable for the active, ongoing treatment of diagnosed inborn errors of amino acid metabolism. Not only are these diseases an emotional hardship for the family, it is also a financial hardship when medical insurance does not cover the costs of these special foods. The estimated cost of supplying special food to a child with PKU is approximately $3000-$10,000/year. However, the cost of caring for a poorly or untreated child with PKU or MSUD and subsequent mental retardation can be tremendously expensive, especially if placement in a group home facility is required. In addition, in the case of MSUD, if the child is not properly treated they will suffer not only mental retardation but also require frequent hospitalizations and intensive care for recurrent metabolic episodes.

The American Academy of Pediatrics feels very strongly that special medical foods that are used in the treatment of amino acid disorders are medical expenses that should be reimbursed. Please see a copy of the American Academy of Pediatrics policy statement regarding the reimbursement for medical foods for inborn errors of metabolism that is attached to my testimony. Treatment with these specialized foods helps prevent mental retardation and possibly death. They should be treated no differently, from a reimbursement standpoint, than life saving or life improving surgery or hospitalization and treatment for multitudes of other medical conditions that are currently single "covered" by medical insurance.

The North Dakota Chapter of the American Academy of Pediatrics feels very strongly that special

medical foods that are used in the treatment of inborn errors of amino acid metabolism are medical expenses that should be reimbursed. We were very pleased when the current law was passed in 1997 that made this a reality for the children in North Dakota. I urge you to help assure that these unfortunate children continue to be treated appropriately and protected from the ravages of mental retardation. This will provide protection for our most precious resources, the children of North Dakota. I would strongly urge you to continue to provide for reimbursement for medical foods for inborn errors of metabolism. I would like to thank you for the opportunity to testify regarding this bill and again I apologize for being unable to attend this committee hearing in person.

If you have any questions, please feel free to contact me at any time. My office number is (701) 780-6110 and my home phone number is (701) 746-9326. Thank you!

American Academy of Pediatrics

Policy Statement

Pediatrics Volume 93, Number 5 May 1994, p 860

Reimbursement for Medical Foods for Inborn Errors of Metabolism (RE9412)

AMERICAN ACADEMY OF PEDIATRICS

Committee on Nutrition

Inborn errors of amino acid metabolism such as phenylketonuria, maternal phenylketonuria, maple syrup urine disease, homocystinuria, methylmalonic acidemia, propionic acidemia, isovaleric acidemia and other disorders of leucine metabolism, glutaric acidemia type I and tyrosinemia types I and II, and urea cycle disorders are rare diseases that are treatable by diet. Treatment might include the restriction of one or more amino acids, the restriction of total nitrogen, or the supplementation of specific substances. Untreated, these diseases culminate in severe mental retardation or death.

Once diagnosis is confirmed, treatment of amino acid and urea cycle disorders must be carefully monitored by a physician with expertise in metabolic diseases. Special medical foods, commercially available, are indispensable for the active, ongoing treatment of diagnosed amino acid and urea cycle disorders. Special medical foods would, if used as the sole dietary source, represent a hazard to affected and healthy children. US Public Law (Publ L) 100-290 defines the term medical food as ". . . a food which is formulated to be consumed or administered enterally under the supervision of a physician and which is intended for the specific dietary management of a disease or condition for which distinctive nutritional requirements, based on recognized scientific principles, are established by medical evaluation."[1]

After passage of Publ L 100-290, many states provided funding for these products through Medicaid, and most states offered assistance through Crippled Children's and Women, Infant, and Children's programs. Some states now have laws mandating private insurance coverage for special medical foods.

It is the position of the American Academy of Pediatrics that special medical foods that are used in the treatment of amino acid and urea cycle disorders are medical expenses that should be reimbursed.

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Ray Yip, MD, MPH, Centers for Disease Control and Prevention

AAP SECTION LIAISON
Ronald M. Lauer, MD, Section on Cardiology

REFERENCE

1. US Congress, 100th Congress. Orphan Drug Amendments of 1988 (Act to Amend the Federal Food, Drug, Cosmetic Act to Revise the Provisions Respecting Orphan Drugs, for Other Purposes). Pub L No. 100-290. Washington, DC: US Government Printing Office.

---------- *The recommendations in this policy statement do not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.*

Return to Contents

My name is Danielle Struxness. ~~and~~ I am a seventeen ~~PKU teenager.~~ years old and a junior in ~~high school~~ at Hazen High School ~~in~~, Hazen North Dakota. I was born in Mobridge ~~SD~~ south Dakota. I was given ~~at~~ new born screening and it was found that I had PKU.

* The main thing on my mind ~~besides it~~ at this age is COLLEGE! I have done alot ~~looking~~ into which ~~college~~ college I would like to attend and what I want to do after I graduate.

~~As far as I have decided so far~~ My decision right now is to attend Minot State University going into Criminal Justice. I would like to work as a probation officer for some experience and then try to find a job as a criminal investigator.

That is what I would like to do. When it comes ~~to~~ finacially, I just don't know. I can borrow around two thousand dollars from the bank. The rest would have to come from scholarships, grants or my parents. I guess you could say I have alot to worry about college.

Now, let's talk about what I have to worry ~~if~~ about just to go on living a normal, healthy every day life.

To take care of my self since I have PKU, I need a metabolic formula to give me all the nutrition that I am missing by not being able to eat being limited to only protein intake. Lots of foods have large amounts of protein in them. Therefore, I am also limited to the foods I eat. To be able to fill up at a meal I also need low protein modified food. With the high prices of metabolic formula and low protein modified food, I need to worry about how my parents and I are ever going to afford to take care of me and go to college. I will be paying for the things I need to take care of myself instead of college.

I guess I could always go to college without being on my low protein diet but then I will have problems learning, listening, and behaving. I will not be able to accomplish everything I could if I could just live a everyday life.

Let's see. Say I made it somehow through college the have the financial aid to take care of myself

while I go to college.. I graduate and find a job. ~~The way I figure my salary to be as a young adult~~ I figure I will not be able to afford to support myself while paying for my nutritional needs. Therefore, I come to you today to ask of you for a small amount of financial aid compared to what I am faced with. Please, allow me to have some insurance that some of my medical needs will taken care of in my many ~~ugly~~ challenging years ahead.

Chairman, members of the committee. My name is Myrna Blackstead, Stacy's mother and what I need to tell you is the importance of a PKU diet.

Over thirty three years ago, when our daughter Stacy was a baby, the only PKU clinic in the Midwest was at the University of Minnesota. It is my understanding that she was their first PKU patient with a normal IQ, and the leading physician in the program was very excited. She has continued to be in that program and has been called back on numerous occasions, to take part in new areas of research for the PKU program.

She had been put on the diet at age 5 1/2 weeks. She was a bubbly, normal toddler except that she was almost always happy. As was encouraged and accepted practice at that time, Stacy went off her diet at age seven. Throughout the rest of her school years, life was not easy for her--low concentration, twice as much work to learn what should have been easy with her mental capacity, low self esteem and immaturity.

She managed to get a general studies degree from a ND University but it had been very hard for her. Her lack of organizational skills kept her from getting the second degree she pursued.

Then in January of 1997, the national PKU newsletter told of the terrible things that were starting to happen to adult PKU's who were off diets. The entire issue was devoted to the importance of the PKU adults getting back on the diet. After reading these warnings, there was no doubt for our family what Stacy had to do. Her immediate reaction was, "I've had my last hamburger."

She had been working full-time at a local daycare and continued to do so. It had looked like this was probably all she could ever do, or deliver pizza as she has also done off and on for years.

But within weeks of going back to a PKU diet, she was a new young women. Her concentration and organizational skills improved within days; she matured to her normal age. She was capable of so much that she hadn't been all these years. And, of course, with all of this, her self-esteem was much improved. Now she is starting her own day care, having done all the planning and organization herself. This would not have been possible when she was off diet.

To be on a PKU diet, she must have a liquid supplement to get all the rest of necessary protein, except the phenylalinine. Last year, using her skills from all of the off-diet years, she made less that $10,000. Her liquid supplement for the year cost in excess of $3,000. In addition she must order her pasta and flour products from a PKU specialty company. Pasta costs are between $2 and $3 per serving.

In September the ND PKU dietitian told her she needs to drink more of the supplement than she had been the previous year. For life.

No matter how much more capable she is of earning a better wage, the cost of keeping this capacity will eat up a large percentage of her income. She really needs the help of her health insurance, which you realize is also costing her a good amount each month. Please make it a law that her insurance company will have to help her keep healthy, permanently, by passing SB2374.

Thank you for your attention do you have any questions?

Good morning

Chairman and members of the committee my name is Stacy Blackstead and I live in Bismarck and have PKU, a metabolic deficiency that does not allow my body to break down one part of protein.

In the almost two years since I went back on my diet, I have better concentration. My outlook on life is better; positive and hopeful. I have higher self esteem. When I do have too much protein, I get down on myself. It is harder to concentrate on details in new situations.

The one thing I notice academically that changed was that my math skills improved. I used to need paper and pen for a simple addition or subtraction problem, but now I can visualize it better in my head or it just comes to me without even thinking.

My drink supplies the nutrients I need to keep a healthy body and mind without the phenylalinine which other wise builds up in my body. The dietary food provides me the same benefit. This supplement to my basic food is expensive but necessary for me to maintain my current level of efficiency.

Stacy Blackstead

Chairman and members of the committee my name is Stacy Blackstead and I live in Bismarck and have PKU. At age seven I was taken off my diet and went back on it at age 31.

In the almost two years since I went back on my diet, I have better concentration. My outlook on life is better; positive and hopeful. I have higher self esteem. When I do have too much protein, I get down on myself. It is harder to concentrate on details in new situations.

The one thing I notice academically that changed was that my math skills improved. I used to need paper and pen for a simple addition or subtraction problem, but now I can visualize it better in my head or it just comes to me without even thinking.

My drink supplies the nutrients I need to keep a healthy body and mind without the phenylalinine which other wise builds up in my body. The dietary food provides me the same benefit. This supplement to my basic food is expensive but necessary for me to maintain my current level of efficiency.

Stacy Blackstead
2124 East Divide
Bismarck

January 7, 1999

To whom it may concern:

My name is David Aakre, I am the proud single parent of four wonderful children. Three of my children are diagnosed with the metabolic disorder called Phenylketonuria (PKU). Namely, Danielle, age 12 Dolan, age 7 and Dalton. age 6. I am very pleased to say I feel my children are developing normally, mentally and physically even though they have this biochemical abnormality which prevents normal brain development if left untreated.

A sincere, and heartfelt thank you needs to be extended for all the people involved in administering the program at the state, and local levels regarding the treatment of this disorder. In this I mean the special medical foods which provide the important nutrients that are an essential part of the food intake pattern of individuals with PKU. The support on an individual basis from a parental perspective provided by the State Health Department has also been extremely helpful in regards to the programs they administer in addition to the special medical foods. However with the winds of budgetary constraints blowing at all government levels, it appears the programs are going to diminish. With this in mind we come to you and ask for your assistance in removing the sunset clause on Senate, Bill 2194, and making it a permanent piece of legislature.

I can remember how devastated I was when I was told our daughter Danielle had tested positive for PKU. Some relief came when I became educated about the disorder, and that it was treatable with a promising outcome of a normal life for her. This has not been an easy task monitoring the children's daily food intake, initiating 504 health plans for school, coordinating menus with daycare, and two schools, and home trying to maintain their required Phenalalinine intake ranging from 500mg for Danielle to 900mg for Dolan. The special medical foods provided by the state have insured the normal growth development of my children. If this program is discontinued, and we the parents of PKU children have to be individually responsible for acquiring these special medical foods some sacrifices may have to be made regarding the children's diet. Don't get me wrong every attempt will be made on my part to insure they continue with their diet, and I will give them everything I am able to, but there would probably be some sacrifices due to the financial situations most of us are in. I have been told the special medical foods which I use i.e. the Maximum XP powder, and Flavonex energy supplement would probably cost in excess of $12,000.00 annually. It is not uncommon for me to spend approximately $500.00 to $600.00 a month at the present time on groceries for the household in order to provide a balanced diet for my children. This on top of other household expenditures would be an extreme financial burden. Your help in this matter will be greatly appreciated.

Getting back to the children, and how their developing and reaching milestones in their lives is a great joy, and also a relief. Knowing their developing normally, and will be able to enjoy normal, healthy, and productive lives is something all the people involved in this program should take credit for, and feel good about. The sense of accomplishment for this type of humanity should be felt by everyone involved, and I am sure all the parents, as well as I, are most grateful. The work and time invested is well worth it when you can watch your Daughter make a break away play and lay-up playing basketball, sing songs at church functions, play trumpet solos at concerts, and helping her study to achieve above average school grades. Playing catch with Dolan and Dalton, teaching them how to ride bike, the list could go on, and to think that the chance for a normal life wouldn't be possible without the dietary treatment is quite heartbreaking.

If anyone would wish to discuss any of this further with me, you may contact me at 701-239-5186 daytime, or 701-428-3797 evenings.

David C. Aakre

David Aakre
P.O. Box 84
220 Spruce St.
Kindred, N.D. 58051

Nancy Ouderkirk

BILL # 2374

My son Evan is now a happy, healthy 3 ½ year old. He's joy, entertainment and fun all rolled up into one little boy. But its taken a lot of hard work by myself and other enlisted professionals to get him where he is today.

The day Evan was born, we didn't spend much time together. After everyone went home, we both fell asleep. And when I awoke, he was gone. The nurse informed me that Evan had been taken to the Neo-Natal Intensive Care unit because he had turned blue and stopped breathing several times that night. There he was attached to a heart and respiratory monitor. At 5 lbs. 14 oz. he already seemed small and he continued to lose weight. Each day that went by he grew more lethargic and would eat less and less. We were force feeding him every three hours. Little did we know that by feeding him this formula, we were only making his situation worse.

After about a week and a half we were allowed to take Evan home with a monitor. We saw his regular doctor who told us that Evan was just fine. I left the doctor's office and returned home. But I hadn't even gotten my jacket off when the doctor himself called me and said that there was an emergency and I had better pack up my son and get him back to the doctor's office. They held the clinic open late for us. And when we arrived, everyone was apologizing to us. Each of the three nurses ushered us back to the doctor's office. Each of them told me that they had never seen one of these before. One of what? In his office, the doctor sat me down and informed me that my son had Phenylketonuria or PKU. I was told that my

son was going to become "dumber" and eventually die with this. Then in passing he mentioned that the dietitian would be calling me. I'm not telling you this to be critical of the doctor, but just to give you an idea of how limited even the physician's knowledge is of PKU.

I then took Evan to the lab for another blood draw to verify the previous test results. As I waited I found that I could no longer contain myself. I was under the impression that there was nothing that I could do. Here I had given birth to this beautiful little boy and he was going to become mentally retarded and die and there wasn't a darn thing I could do but sit back and watch the whole awful scenario slowly unfold.

I taught public school for over ten years. I've seen how students and adults treat people who are different then they are. I've seen how cruel they are to mentally handicapped people. I remember thinking that if he really had to die why couldn't it be over soon rather than make him suffer.

Later that evening the dietitian, Cathy Breedon, called me. After what I had heard, it took her a long time to convince me that my child was going to be O.K. I just needed to feed him properly. Once I got past the words "mentally retarded" I was fine. But what about Evan? His lethargic condition was getting worse by the day. Being PKU children are so rare, meeting their needs immediately was extremely difficult. We were coming into a weekend and Cathy couldn't find the special formula that Evan so desperately needed. She was finally able to locate a can of the toddler/adult formula and with some modifications we made a suitable substitute until the proper formula arrived.

Once Evan began drinking the correct formula his condition changed dramatically! His breathing got better. He was more alert. He slept and ate better. He started to put on and hold weight. His phenylalanine level quickly came down to a manageable point. Evan was a completely different child.

As Evan has gotten older he's had problems with chronic ear infections. He's also had a reoccurring upper respiratory infection. In most cases one would take their child to the doctor, receive medication, go home and wait for it to work. Every time Evan gets a cold or ear infection, it affects his blood levels. Like anyone else, when Evan's sick his body breaks down additional body proteins to fight the infection. Protein, no matter where it comes from, contains phenylalanine which is toxic to his brain. To compound the problem, because of the rarity of the condition, I have to be the expert when we go to the emergency room or visit any doctor other than his own. We have a difficult time finding medication for Evan that doesn't contain Nutrasweet or Aspartame. Nutrasweet or Aspartame is straight phenylalanine and he can't have that. I had to deal with a doctor in one instance who insisted that there was not much Nutrasweet in this particular medication. I gave up on him and talked to the pharmacist who is a friend of mine. I asked Dave just how much PHE was in each teaspoon. He said 26 mgs. Evan needed two teaspoons per day for 2 weeks. 52 mgs was equal to an entire meal for him at that time. How do you ask your child, "Would you like to eat this meal or would you like to take your medicine?"

When PKU children experience high levels of PHE it can effect their behavior as well as many other things. One night Evan couldn't stop coughing so I took him

to the emergency room. The doctor informed me that the treatment for this crupy type of cough was a steroid shot. He said that it wouldn't hurt Evan and since I had little prior knowledge of the effects of steroids I gave my consent. It wasn't long before Evan was the most difficult baby on the face of the earth! He was kicking, screaming, biting, hitting, etc. This type of behavior went on for days and he smelled terrible. (Phenylalanine has a sickening sort of musty odor.) I didn't know what had happened, but after talking with Evan's doctor I learned that the steroid shot had set him off. Steroids break down proteins. I didn't know and the ER doctor didn't make the connection that protein is a PKU child's problem. Being it was a long acting shot, it was almost two weeks for Evan to get back to normal behavior. It's just like going through withdrawals. I've learned a lot during the past three years.

Individuals with PKU are very strict vegetarians, not by choice, but out of necessity. They also may eat many specially prepared foods that may be ordered through four different companies. It's great that these items are available, however, they don't come without cost. The cost of feeding a maturing PKU child is estimated at $10,000 per year. Thanks to the State Health Department our children have received the necessary formula which is 90% of their total diet. There is no other replacement for the formula. Without this formula, phenylalanine will build up to a toxic level in their brains. No other food will save our children. Without their formula it's only a matter of time before each one of the children you see before you today will become mentally retarded and institutionalized. As of June 1999, the grant will run out which means an additional $5,000 per year added to our budgets. (To purchase this formula through the hospital is $50 per can.) This is over half of my

income just to feed Evan. That's not taking into consideration that he needs clothing and a home or the fact that my income must meet his sister's and my needs as well. In the case of Cole Johnson with MSUD, if he doesn't get his formula, he dies. There are other programs available to help defray the cost of formula, however, many of us make too much to qualify but not enough to support our children. I'm not being melodramatic, I'm simply stating the facts.

With your help our children can become healthy, valued, contributing, not to mention tax paying members of our communities. Without it our children could very likely become wards of the state. Just one PKU child being supported in a state institution will cost much more than a whole year of support for all 22 PKU residents currently in our state. If this necessary formula and food is not provided to our children the result is mental retardation. This is a price far too great for any individual or family to have to pay. This expenditure for third party payer is far less than the cost of letting one person suffer the consequences of untreated PKU both in human and financial terms. The mental retardation that results is profound, costly, unnecessary, irreversible and inhumane.

The state of North Dakota mandates the screening for PKU children at birth because it can be successfully treated. Treatment is costly. And to help cover treatment cost, we purchase insurance. In **most** cases the insurance companies may be correct in citing their policy that food is not a medical expense. However, in the case of PKU, the formula and the food are the medications. It's the **only** treatment for this condition. Many parents boast about their children being above normal. We as PKU parents boast of how normal our kids are! Treatment for PKU

allows our children to lead a normal life. We need the help of the insurance companies and the state. And we need your help to give our children of North Dakota that chance at a normal life. Our kids our counting on you.

On behalf of my son Evan, his sister/my daughter Kailynn, and myself, we'd sincerely like to thank you for your consideration.

I'm writing this testimony in support of Senate Bill number 2374

I don't really understand why I should need to do this as I am quite confused as to why insurance companies have not had to provide reimbursement for low protein modified food products and metabolic formula, which are specially formulated, to be used in the treatment of individuals who have an inherited metabolic disease and prescribed by a physician. The only treatment for Phenylketonuria is dietary. The result of not complying with a specialized rigid diet and keeping phenylalanine blood levels within allowable limits is brain damage.

Instead of spending time, energy and money fighting for this bill, we should be commending all the individuals who are afflicted with this disease for having the courage to stay on this rigid diet. I would venture to guess most of us have been on a diet at least once in their lifetime, you know, to lose a few extra pounds. How easy was it for you? It's hard to follow a diet, and if you choose not to follow it one day or one week perhaps around the holidays, you knew you would get back on track sometime. Well, these individuals don't have that choice. In order to limit any damage they might sustain, they need to follow this diet every day of their life. That's right, every day of their life. I don't know about you, but I'm glad I don't have to comply to these standards.

Now let's talk about food choices. When we go on a diet, we have plenty of food choices. After all, we have four basic food groups. We might not think so when we are dieting, but there are lots of choices within these food groups. Individuals with PKU aren't as fortunate as we are. They don't have the luxury of four basic food groups. Their diet is extremely rigid with few food choices. Yet the individuals with this disease tackle this diet with courage. Yes, it is difficult, but they don't have the option of quitting if they want to lead a "normal" life and be a functioning member of this state -- one who can get and keep a job, pay taxes, pay for insurance, etc.

Every baby is screened for this disease when they are born in this state. Aren't we fortunate to have this valuable information, that our child has PKU, so we can help our children prevent mental retardation. Yes, that's right, mental retardation. Before this screening was initiated, there were individuals who had PKU who are now mentally retarded and living in group homes. They never had the chance or choice to go on this specialized diet. Now we have this information, know how to treat it, but cannot afford to as it currently costs $4,000 to $6,000 for just the metabolic formula for one child a year, and insurances in this state will not assist in covering this expense.

So, you see, I am really questioning why this is even an issue with insurance companies. On January 12, 1999, I called my insurance company to find out some information. I inquired as to if my policy would provide coverage for Viagra if a doctor prescribed it. The answer was yes, they would. Now, I ask you, can you get and keep a job, have your own place to live, pay taxes, pay for insurance, etc., without Viagra.

I also asked if they would cover diet pills if a doctor prescribed it. The answer was if there was enough documentation that this treatment was warranted, they would. So, all you need to obtain diet pills is: 1) a physician's prescription, and 2) documentation that this treatment is warranted.

Then I asked the individual who was representing the company if my Blue Cross/Blue Shield policy covered treatment for PKU, and she informed me that they would only cover the testing.

Let's see, they'll cover Viagra and diet pills, but they won't cover low protein modified food products or metabolic formula for the treatment of an inherited metabolic disease. Does this make sense to you?

A person diagnosed with PKU has documentation that this is an inherited metabolic disease which results in brain damage if untreated, and the metabolic formula has to be prescribed. What else do the insurance companies need?

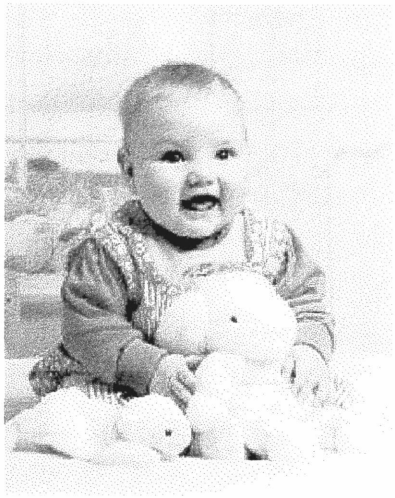
After receiving this information, I became even more frustrated and confused. How could this be? An individual who has been diagnosed with phenylketonuria for which the only treatment is a low protein diet and can result in brain damage and mental retardation, is not allowed to receive reimbursement for expenses incurred to treat this inherited metabolic disease.

I worry about my son as he becomes an independent adult. How is he going to afford insurance coverage much less the cost of his metabolic formula when his insurance company will not assist in the purchase of this formula. He has a strong desire to obtain a job and become independent, but without the insurance company assisting in obtaining his metabolic formula he will not be able to stay on his diet because he will have normal living expenses as we all do.

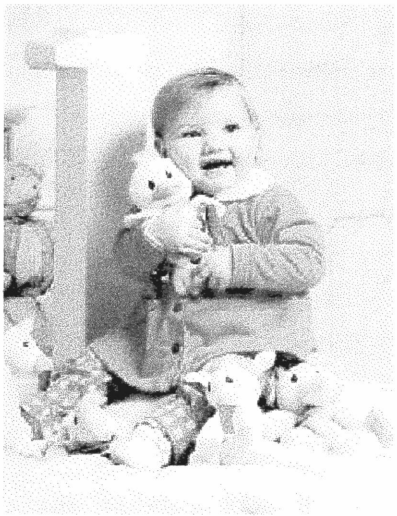
Thank you for listening to my concerns. I hope you will support Senate Bill 2374.

Gayle Heskin

Gayle Heskin



Rebekah Wangler
6 mo. old



Rebekah Wangler
1 yr. old

To Whom It May Concern,

I am writing this testimonial in support of Senate Bill # 2374, mandating insurance coverage for metabolic formula and low protein modified foods.

Our daughter, Rebekah Kathryn Wangler, was born October 28, 1998, and was immediately diagnosed with the very rare metabolic disease of Phenylketonuria. In case you are unaware of, like we were, the number 1 concern of metabolic disorders is mental retardation. Instead of being upset and angry, my husband and I could only focus on how happy and relieved we were that there was: 1.) Mandatory tests available to detect PKU: 2.) A doctor (Dr. Alan Kenien) in the state who specializes in metabolic disorders; 3.) A dietician (Kathy Breedon) to watch over and supervise Rebekah's very limited and lifelong diet; and finally, 4.) That Rebekah does have a chance to live a very normal and healthy life, something that years ago, was not a reality.

Just as it is required by state law for hospitals to perform mandatory testing on all newborns for PKU, so should it be mandatory for insurances to provide coverage on metabolic formula and low protein modified foods. Unfortunately, Rebekah does not have a choice of which foods she can eat. She must be on a lifelong diet of metabolic formula and low protein modified foods in order to live a normal and healthy life. The means are available for this to happen, it is the financial support that is needed. By not mandating insurances to provide financial support, these children face the grim reality of becoming mentally retarded, which, in the long run, will be more costly for our state. Other states require mandatory coverage for the metabolic formula and low protein modified foods. What a sad shame it would be if our families and children were forced to leave this beautiful state of North Dakota because they cannot receive this coverage that is financially needed. The increasing cost of metabolic formula is of great concern. To date, a case of Phenex 2 (6 cans) which is the metabolic formula Rebekah is currently taking under the direction of her physician, costs $160.00. She currently goes through approximately a case a month. Soon, we

will have to start purchasing her low protein modified foods which will cost another $200.00-$300.00 a month. Our current financial situation will not allow us to cover these increasing monthly costs.

It is my prayer that you realize how important it is to mandate insurance coverage for the future of our children. As I listen to speeches by our president, senators, governors, and congressman, they constantly mention how we must support certain bills and programs for our children's future. By mandating insurance coverage of metabolic formula and low protein modified foods, you can put your words into action. I mentioned earlier that those children born with inherited metabolic disorders have a chance to live normal and healthy lives. That chance depends highly on the support of Senate Bill #2374. The first step has been taken in helping our children lead normal and healthy lives by making the PKU test mandatory, now I pray that you will take the next step in making insurance coverage mandatory to insure our children the chance to continue their normal lives.

SINCERELY,

Dale & Kris Wangler
Rugby, N.D.

Senate Bill 2374

PKU! I remember my new baby's doctor saying those 3 letters to me when she was 7 days old. It meant absolutely nothing to me at that point. All I was hearing was "your baby is sick and she is not normal."

Although now I know by just looking at her that is only partially true. She is better than "normal", she is **miracle** and that was made possible by just eating the right food!

Kaylin is now 2 ½ years old and she loves her metabolic formula. We thank God everyday that she drinks all of her metabolic formula, which is the key to maintaining her health.

My husband and I were very troubled to find out that the state may no longer be able to afford the cost of the metabolic formula that Kaylin drinks. My husband and I both work but our combined income is still relatively low so our only hope is that with mandated insurance coverage, our daughter will still be able to get the metabolic formula and low protein modified foods that she so desperately needs to maintain her health.

As you probably already know, the alternative to her not receiving her metabolic formula and low protein modified foods is **permanent, irreversible mental retardation.**

For now, we continue to weigh and measure everything that Kaylin eats and we keep track of the amount of phe she has eaten each day. Kaylin has begun to learn what foods are acceptable for her to eat and what is not. The terms we use if my husband and I are eating ice cream for example and she asks if she can have some is "it is not your kind" then upon giving her a scoop of sorbet telling her "this is your kind". It is not always as easy as it sounds, recently it has been hard for her since she is now at an age where she is wanting to discover and try new things.

Kaylin's least favorite part of PKU are her weekly blood draws and every 3 months she must have about 4 tubes of blood drawn for her Metabolic clinics. She remembers her past experiences every time that we go to the lab and begins to cry as we walk in the door. She has also asked me on several occasions of going to the doctor for checkups, "is doctor going to take all my blood again, it hurts" as her eyes well up with tears.

We are asking that you will please support mandated insurance coverage for our daughter and all the other beautiful, healthy PKU children of this state. Thank you.

Marshall, Tammy and Kaylin Anderson
Manvel, ND

Kim and Brenda Ankenbauer

Kim and Brenda Ankenbauer
1108 Main Avenue
Washburn, ND 58577

RE: Senate Bill # 2374
Mandating Insurance Coverage for Metabolic Formula and Low Protein modified foods for PKU and NSD

Dear Sirs;

Kim and I are the parents of three girls. Torie 15, Tera 11, and Candace 6. Kim works at the Falkirk Mine near Washburn, and I work at the local bank part-time. When our middle child was born with PKU, we were totally in the dark. We had never heard of PKU, which I'm sure alot of you have never heard of it either. Well, believe me we learnt fast. The first thing we heard a very strict diet will need to be followed or the child could become severely retarded. If you have children, you can imagine the fear we felt. We were willing to do whatever it took to make sure Tera would grow up to be a healthy little girl . First thing we had to do was to get the right formula. Like most new mothers, it was a trip to your local drug store or super market to pick up formula. Tera's Metabolic formula that she needed , could not be bought there. We learned fast that it was funded by the State, and that we would have to pick it up at the State Capital, or it could be shipped to our home. It was very hard at first to get Tera to drink it. She hated it. We new it was very important that she drink it, and never gave up trying. When she became old enough for foods to be introduced, it became a whole new ball game. Everything she put into her mouth had to be carefully measured or weighed, as Tera was missing an enzyme to break up an amino acid called phenylalanine hydroxylase. So basically, we had to feed her only what her little body could use. This was carefully monitored through blood tests to make sure she was not getting to much. It became alittle bit harder, because we found that just about every food had phenylalanine in it. Some foods such as meat, cheese, and in Tera's case even a slice of bread was a no no. We were about pulling our hair out as to what we were going to feed this child. We found out, through Tera's doctors, and PKU team that you could get low protein modified foods , and a number of other products from a Dietary Specialty place out of New York. They told us it would be expensive, and it was, but we had to have something to feed her. We soon learned to make bread. See we can't go to the store to buy a loaf of bread for her. She couldn't have the bread we eat. As Tera grew older, she still needed the special milk, which thank God was being paid for by the State, but the food we have always continued to by for her through "Dietary Specialties". Her whole grocery list is not bought there, but the things that we just can't by for her in your local grocery store is. The rest of the family does not use any of these products. We think of it as Tera's medication. See in a way it is. It's not much different then being a diabetic and needing insulin. The products are very expensive and now we found out the State is not funding the formula either. The formula is so important to these kids. It is all the vitamins and minerals that they are not getting, because of such a strict medical diet. We are really concerned as parents as how we will pay for all this, and it just doesn't seem fair that it's all medical and they won't help us. We pay dearly for premiums monthly. Why can't we get some help for Tera's needs. We worry about down the road, when Tera is on her own. This is a lifetime diet, how as a young adult can she afford her medical needs. This disease is so easy to control with proper management of diet. At first when we heard about the diet, we couldn't believe a diet could be so important. We asked the question " What

kind of medicine does she have to take". Of course there is no medications, her Metabolic formula and special diet is her medication. Tera is as normal as any 11 year old girl. She is in Girl Scouts, plays piano and flute, loves to run in the track programs. She likes to go camping, boating, and swimming in the summer. She is on the girls 5th and 6th grade basketball team. She carries her lunch to school every day, as it is her special lunch and still drinks her Metabolic formula daily. She goes to birthday parties and takes her own cake. (made by mom with the special flour we order). If it's a pizza party, mom makes her a special pizza. She can't have regular pizza. Tera does well on her diet and excepts it well. She doesn't know any different. With all the added expense of her diet, I hate to have to tell her we can't afford to buy your medicine this month. That's just not fair. Please consider mandating insurance coverage for PKU Metabolic Formula and Low protein modified foods for PKU and NSD, as these kids are worth it! Thank-you for taking the time to read our letter.

Sincerely,

Kim , Brenda, Torie, Tera and Candace Ankenbauer

To Who It May Concern

Ref: Support Senate Bill 2374.

I'm writing on behalf on my 4 year old daughter, Carolynn Wollenberg. Carolynn was born on July 15, 1994 with PKU (phenylketonuria) and has to stay on a low protein DIET FOR LIFE. This diet contains modified low protein foods and a metabolic formula. The formula alone costs about $158.00 for 6 cans.

If she doesn't maintain her diet, she will end up having many problems as she goes through life. She'll be very irritable, hard to live with, slow, have poor grades in school, and have behavioral problems that could lead to criminal acts, difficulty holding a job to support herself and she might not be able to obtain a drivers license. This could lead to having someone else take care of her the rest of her life.

So please won't you help us to mandate insurance coverage for the metabolic formulas and the

modified low protein food. Lets take off the 2 year trial period and make it a law.

Thank you for taking the time to read this.

Sincerely yours
Jerry Wollenberg
Mother of Carolynn Wollenberg
9652 County Rd 4
Hamilton ND 58238-9716

1-11-99

To whom it may concern,

We are writing you today to express our concern regarding the Bill # 2374 regarding mandated insurance nutrition coverage for children with metabolic disorders. As you know one of the greatest resources in North Dakota are our children, it does not make sense to cut programs that benefit them and their health. We would appreciate your careful consideration regarding this matter. Enclosed is a picture of our son who has PKU. We hope all the children with metabolic disorders will be as healthy as our son.

Sincerely

Keith and Linda Ulmer

Keith and Linda Ulmer

To whom it may concern, Ref: State bill number 2374 Page 1

My name is Amy Johnson,

I will try to keep this short and simple.

Our son was born with a rare genectic disease called MSUD (Maple Syrup Urine Disease). It affects about one in 300,000 births. This disease affects the way the body processes certain components of protein. Including the amino acids Leucine, Isoleucine and Valine. Children with this disorder lack an enzyme that helps break up the protein, when the protein components accumulate in the blood, they cause a toxic effect that interfers with brain function. If the disease goes untreated, or the child doesn't drink his special formula, symptoms can progress to seizures, coma and death. The disease must be caught quickly at birth, If not the child usally dies by 14 days of age. Cole was a very lucky baby he was in a coma for 5 days, before being diagnosed at 16 days of age. Dr's didn't know if he would have irreversible brain damage. But we have been blessed. Cole functions like any normal 7 year old. Almost any illness Cole suffers can cause a metabolic imbalance, which can be life-threatening.

He has been Hospitalized 6 times. The most serious was about 3 years ago. Cole had the flu, and was not drinking his formula. After acouple of days Dr. Kenien put him in the Hospital. For his normal

glucose IV. But after acouple of hours he started having seizures and went into a metabolic crisis with severe brain edema. Thanks to the quick work of Dr. Kenien and Dr. Morton from Lancator, Penn.

Cole survived. Cole Needs his special formula and foods to survive, and without them, We would not have our son. Without the help of the ND MCH Division, formula for Cole will cost us $7,325.00 a year . Plus the cost of low protein modified foods. That would cost us at least $610.00 A Month. How would you like to live with the feeling, if you can't make you sons formula payment, he will Die.

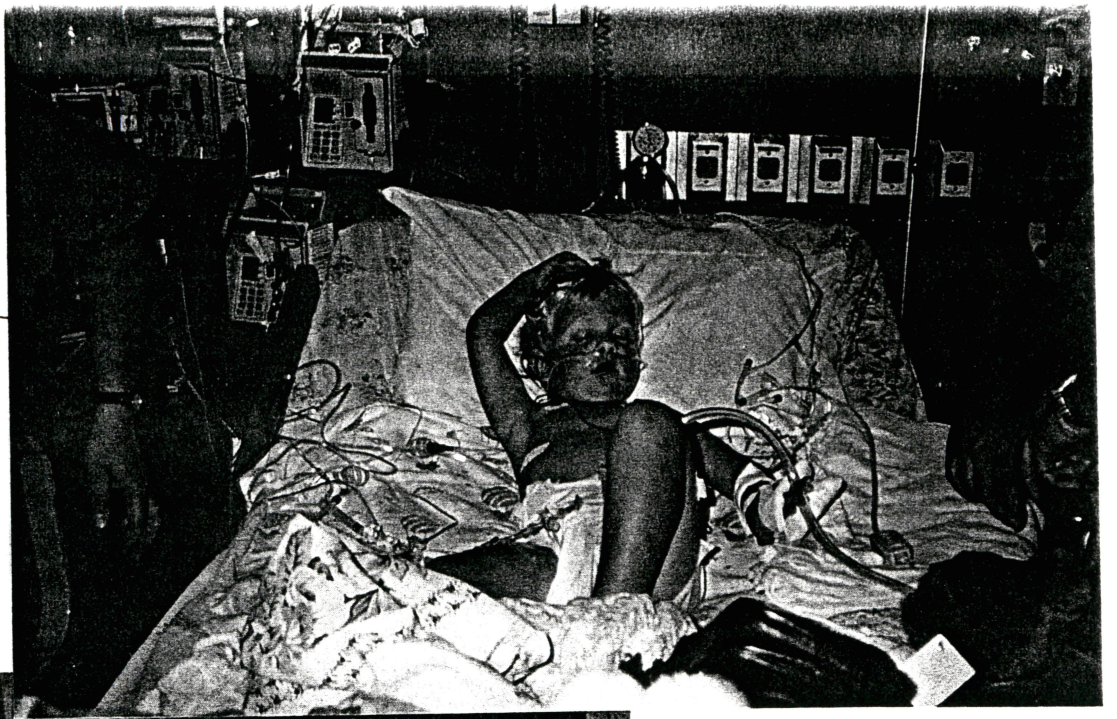
Thank You

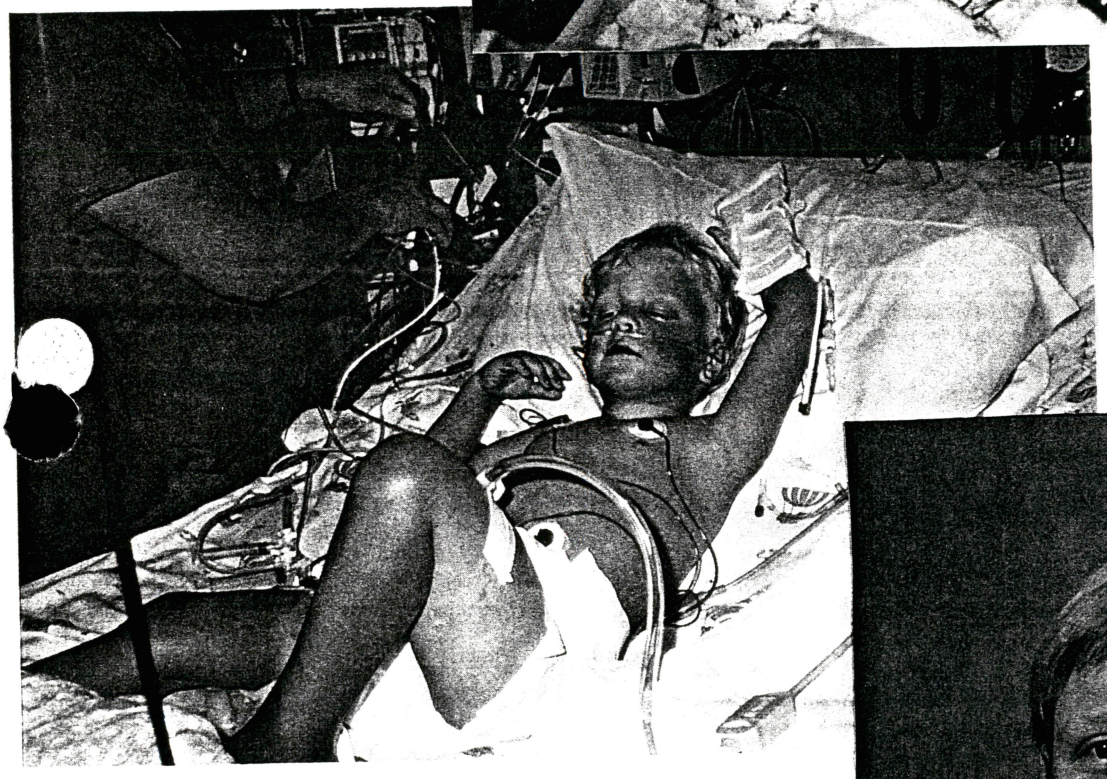
Sincerly

Amy & Jeff Johnson

This is
What happens
to our son Cole,
when he doesn't
get his formula

1.) metabolic
crisis

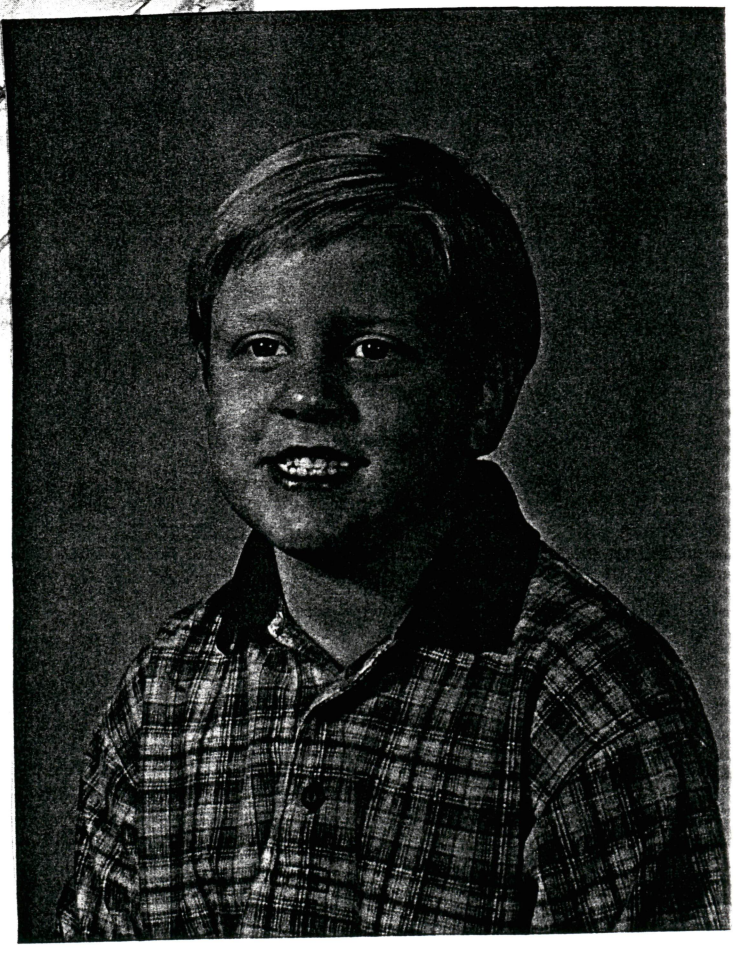




2.) Coma

3.) Death.

This is Cole School
Picture this year.
He drinks his
Formula everyday.



Cole 7 yrs old

TESTIMONY FOR SB 2374

Before the House Human Service Committee
Prepared by Senator Judy Lee

March 8, 1999

SB 2374 will continue the program which provides payment for special formula and food products for citizens with certain metabolic diseases: phenylketonuria, or PKU; and maple syrup urine disease, or MSUD.

These two diseases are quite rare - inherited metabolic diseases in which ordinary food is not processed in a normal fashion by the person with the disease. Screening of newborns has been done since the late 1960's for PKU, which occurs in 1/10,000 births. MSUD occurs in 1/200,000 births. ND currently has 18 individuals who have these diseases, 17 with PKU and 1 with MSUD.

If undetected, the persons with these diseases will be developmentally disabled and require lifetime assistance. If discovered promptly, the special formulas can be prescribed and the children will grow to have normal intelligence and lead normal and productive lives. Up until recently, medical practice was to discontinue the special foods after the child reached his/her teenage years, because it was no longer considered necessary. Recent research and experience has clearly demonstrated that, for at least some of the patients, lifelong use of the special foods is necessary in order to maintain a normal life.

Formula for the PKU children costs approximately $3,800 per year. Low protein modified food products cost from $1,500-$2,500. Formula costs for MSUD are $7,000 per year. The Department of Health currently provides special formulas for these individuals. Current law permits the Health Department to use Maternal & Child Health (MCH) block grant funds to provide benefits only to age 22 as well as for PKU women considered to be of childbearing age up to age 44.

Two years ago we passed legislation which called for insurance coverage of some of these costs. Information gathered since then indicates that only about half of the families are covered by insurance policies which include these expensive food products.

The Senate Human Services Committee worked with other interested parties to find a way to assist these individuals with the high cost of these products which are necessary to maintain a normal life. SB 2374 provides for formula and medically needed food for persons up to age 22, and for women up to age 44 will

be provided by MCH grants through the Health Department, unless the persons are eligible for Medicaid. If the person is Medicaid eligible, the Department of Human Services will cover the cost of formula and food products. If food is not available under Medicaid coverage, the Health Department will cover the cost. Insurance will cover the cost for those over the eligibility income and for males over age 22.

Our committee's goal was to find a way to provide these products for all of the 18 individuals now needing them and not exclude those whose insurance would not cover it. This bill does a good job of providing support for these families who need these expensive food products and formula in order to keep their children healthy and prevent mental retardation.

Amendment:

The amendment to SB 2374 says that insurance coverage will be required for the special food products and formula needed for PKU and MSUD patients, if they are ineligible for programs from the Health Department or Department of Human Services.

TESTIMONY BEFORE THE HOUSE HUMAN SERVICES COMMITTEE
REGARDING SENATE BILL 2374
MARCH 8, 1999

Chairman Price, members of the Committee, I am David Zentner, Director of Medical Services for the Department of Human Services. I appear before you today to provide information and to recommend an amendment to this bill.

The North Dakota Medicaid Program currently pays for the cost of formula for individuals with PKU or maple syrup urine disease and we intend to continue coverage in the future. Payment is made on behalf of any eligible recipient based on medical necessity relating to the nutritional requirements of individuals with inherited metabolic disease. The department does require prior authorization before payment can be authorized. Payment is made to enrolled medical providers based on a written prescription from a physician after review and approval by our medical consultant.

The Department does have concern with Section 1, paragraph 4 of this bill. The language implies that medical benefits coverage for insurance companies would not be liable for payment to the extent those benefits are available to an individual under a department of human services programs.

The Medicaid Program is considered the payor of last resort. Federal Medicaid law prohibits any health insurer from taking into account that the individual is eligible for or is provided medical assistance under the Medicaid plan for any state. Therefore, if a Medicaid recipient has other health insurance coverage, payment must first be sought and obtained from that source before Medicaid can pay for any services. Not following these requirements could jeopardize federal Medicaid funding.

The current language conflicts with the above requirement because it relieves responsibility to an insurance company for services covered under this bill, if coverage is available through a Department of Human Services Program, which includes Medicaid. The Department suggests an amendment on lines 13 and 14 of page 2, removing "or under a department of human services program."

I would be happy to answer any questions you may have.

Testimony
on
SB 2374, PKU Insurance Benefits
before the
House Human Services Committee
by
Murray G. Sagsveen, Department of Health

March 8, 1999

Good morning Rep. Price and members of the Committee. I am Murray G. Sagsveen, State Health Officer. I am pleased to present testimony regarding the Department's PKU screening and special formula food program.

Phenylketonuria (PKU) and Maple Syrup Urine Disease (MSUD) are two uncommon inherited metabolic diseases. The North Dakota Department of Health has conducted a metabolic screening program for PKU and provided special formula to PKU children since the late 1960s. Children with PKU, if detected at an early stage, may grow to have normal intelligence if a moderately expensive dietary treatment is provided.

Phenylketonuria occurs in 1/10,000 births. Maple Syrup Urine Disease occurs in 1/200,000 births. North Dakota has approximately 8,500 births a year. Thus, on average, one child with PKU is born in North Dakota every 14 months. Formula costs for children with PKU who stay continuously on the formula are approximately $3,800 per year. Low protein modified food costs range from $1,500 to $2,500 per year according to information from other states. Formula costs for MSUD are $7,000 per year. The Department of Health currently provides special formula for 17 children (under the age of 22) with PKU and one child with MSUD. Approximately $100,000 of Maternal and Child Health [MCH] block grant funds will be used this biennium to purchase the needed *formulas.*

The Department of Health also collaborates with the Department of Human Services, Children's Special Health Services (CSHS) Division, formerly Crippled Children's Services, in conducting metabolic specialty clinics in North Dakota. These clinics provide multispecialty services funded with CSHS monies.

While special formula and low protein modified food products were previously recommended for children with PKU through preschool and the early elementary years when commencement of a regular diet would begin, some experts now recommend life-long treatment of PKU with these products. Under current law, the Department is authorized to use MCH block grant funds to provide the benefits only to PKU children up to age 22, as well as pregnant PKU women (or those seeking to become pregnant up to age 44).

The State of North Dakota has for three decades required that children be screened for PKU. Experience has shown that nutritional therapy can effectively prevent severe mental retardation in children with PKU. For that reason, the Department of Health has provided formula and low-protein special foods that have helped many families and children over the years.

There are three additional issues I would like to address. The first is the Senate amendment to the bill. As the result of an internal Department of Health review of the metabolic food program and additional questions raised during the Senate hearings focusing on the purpose of the Department's Program -- which is to avoid the cost and burden of severe mental retardation -- the Senate adopted an amendment providing that a health insurance policy is not required to cover --

"[P]rotein modified food products or medical fluid for an individual to the extent that those benefits are available to that individual under a department of health program . . .."

From a Department of Health perspective, the amendment is significant for several reasons:

- The Department will be able to maintain a register and assure that all eligible individuals, particularly children, receive necessary formula and low-protein food products. If insurance companies had this responsibility, those with a metabolic condition could be subject to high deductibles or co-payments, and the Department would not have the same ability to monitor the use of formula and special food products by these individuals to assure that they are receiving the appropriate amount of these products (which varies based on the age and weight of the individual).
- Individuals would move on and off health insurance coverage or change policies under which they may have different co-payments and deductibles that could disrupt the continuity of these special dietary supplements.
- From a public health and budgetary perspective, the Senate concluded it is desirable to adopt a policy that would maintain the health of an individual with a metabolic condition and avoid the risk of mental retardation.

The second issue relates to an amendment the Department of Human Services will offer that removes a reference to their programs because of a concern that this language could be construed as being in conflict with the state Medicaid plan. Because Medicaid already distributes formula to Medicaid beneficiaries needing this treatment, we have no objection to their amendment.

The third issue concerns a requirement set forth in section 25-17-03 (2) that defines an individual eligible for PKU formula benefits as a person in a family that "is unable to pay the cost of such treatment." It appears that the Department of Health has never issued any rules to

objectively define this section. Rather than attempt to define it in this legislation, which relates to the repeal of a "sunset" provision in an insurance mandate, we will address this matter in a rulemaking proceeding that we will initiate as soon as the 56th Legislative Assembly adjourns.

* * *

I will answer any questions that you or other members of the committee may have about the Department's metabolic food program.

# # #

My name is Anne Christians I was born and raised in Fargo. I am 22 years old and a senior at NDSU. I am also a phenylketonuric. This means I have PKU. This is not so much what I have as who I am. I have been on a restricted diet my entire life as I will be forever. I am lacking an enzyme in my liver that processes a certain protein. Therefore, I am a strict vegetarian. I eat no meat, dairy products, eggs, or fish. I can't even drink diet soda. I drink a special formula to provide my body with all the nutrients that I can't get through my diet. This formula is what has guaranteed me a "normal life". Without it I would have mental retardation. I feel very fortunate that I was born after PKU was diagnosed and a treatment had been developed. However, this treatment does not end. For me to maintain and continue my life as a successful student and productive member of society I need this formula. It has been documented in adults that without this diet there can be neurological problems even to the point of becoming psychotic. My thought processes would become unclear and I would become lethargic and unmotivated. Physically , I could have seizures, headaches and sleep disorders. It has also been noted that adults who have PKU and are off the diet have a much higher incidence of criminal behavior and prison sentences.

As a woman, this is an especially important issue to me, as I will also need this formula to ensure the health of my unborn children. If I were not on this diet I would have elevated levels of Phenylalanine in my blood. As the baby would get its nourishment through my bloodstream, the child would be born with mental retardation, even though it would not have PKU. I will need to be on this diet, with the formula, throughout my child bearing years. On the diet I will be able to have normal healthy babies.

At the present time I am completing a one year internship in Medical Technology

at Trinity Medical Center in Minot. After this summer I will be looking for a job. Please don't force me to have to leave the State of North Dakota and seek employment in a state that does cover PKU. As I am now 22, I no longer fall under the Maternal and Child Health Department guidelines. The state will no longer provide me with this formula. If the insurance coverage is allowed expire it would be difficult for me to afford this cost. In my case, this formula costs about $5000 a year. This a genetic disorder that I did not ask for, nor could I control being born with it. Please do not penalize me for this. You hold the quality of my life in your hands.

There are so many other things that I could say. I feel very lucky when I think about my life and all the wonderful things I've been able to see and do compared to how my life would have been without this diet, my formula and the people who care. On behalf of myself, my brother, who also has PKU, and all the other PKU children in the state I urge you to remove the sunset clause from Senate Bill No. 2374.

Dear Chairman, and members of the Committee.

My name is David Aakre, I am the proud single parent of four wonderful children. Three of my children are diagnosed with the metabolic disorder called Phenylketonuria (PKU). Namely, Danielle, age 12, Dolan, age 7 and Dalton, age 6. I am very pleased to say I feel my children are developing normally mentally, and physically even though they have this biochemical abnormality which prevents normal brain development if left untreated. PKU results from a deficiency of the enzyme responsible for metabolizing the amino acid Phenylalanine. This results in the build-up of Phenylalanine to toxic levels within their body. If untreated the children will suffer irreversible brain damage as well as severe and progressive neurological disorders. We treat PKU by restricting the children's protein intake, and supplementing their diet with a metabolic formula that has all the Phenylalanine removed. This formula provides calories, and amino acids (excluding Phenylalanine). Without it a PKU patient would experience severe malnutrition leading to failure-to-thrive, and in some cases death.

A sincere, and heartfelt thank you needs to be extended for all the people that have been involved in administering the treatment at the state, and local levels for this disorder. In this I mean the special medical foods which provide the important nutrients that are an essential part of the food intake pattern of individuals with PKU. The support on an individual basis from a parental perspective provided by the State Health Department has also been extremely helpful in regards to the programs they administer in addition to the special medical foods. However with the winds of budgetary constraints blowing at all government levels, it appears the programs are going to diminish. With this in mind we come to you and ask for your assistance in removing the sunset clause on Senate, Bill 2194, and voting favorably for Senate Bill 2374 making it a permanent piece of legislature.

You see we have three options. Number one we could discontinue treating our children for PKU, and watch their mental capacity to function in daily living slowly deteriorate followed by an onslaught of a number of neurological disorders. I couldn't imagine any loving parent taking this course of action.

Number two, we could continue treating PKU the best way we can by restricting the children's protein intake, without supplementing with low protein metabolic foods. This treatment would allow us to slowly watch our children suffer from malnutrition, and basically wither away. Another unacceptable course of action.

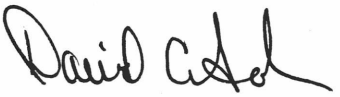
Number three, we could continue with the same treatment level we are currently using but at a severe financial burden. This of course is what I will be choosing to do because all other options would be unbearable.

I can remember how devastated I was when I was told our daughter Danielle had tested positive for PKU. Some relief came when I became educated about the disorder, and that it was treatable with a promising outcome of a normal life for her. This has not been an easy task monitoring the children's daily

food intake by recording and calculating everything they eat to keep track of the prescribed amount of Phenylalanine for each of the children with PKU every day. Initiating 504 health plans for school, Individual Educational Plans, coordinating menus with daycare, and two schools, and home trying to maintain their required Phenylalanine intake ranging from 500mg for Danielle to 900mg for Dolan. The special medical foods provided by the state have insured the normal growth development of my children. If this program is discontinued, and we the parents of PKU children have to be individually responsible for acquiring these special medical foods without any help some major changes in our lives will occur due to the financial situations most of us are in. The special medical foods which I use, the Maximum XP powder, and Flavonex energy supplement would cost in excess of $1,100.00 monthly. This is going to consume more than 1/3 of my household income, I'm not sure as to what's going to happen to us, or anyone else for that matter. I don't think there are too many people who can recover from a sudden debt load like this. It is not uncommon for me to spend approximately $500.00 to $600.00 a month at the present time on groceries for the household in order to provide a balanced diet for my children. This on top of other household expenditures would be an extreme financial burden. Your help in this matter will be greatly appreciated.

Getting back to the children, and how their developing and reaching milestones in their lives is a great joy, and also a relief. Knowing their developing normally, and will be able to enjoy normal, healthy, and productive lives is something all the people involved should take credit for, and feel good about. The sense of accomplishment for this type of humanity should be felt by everyone involved, and I am sure all the parents, as well as I, are most grateful. The work and time invested is well worth it when you can watch your Daughter make a break away play and lay-up playing basketball, sing songs at church functions, play trumpet solos at concerts, and helping her study to achieve above average school grades. Playing catch with Dolan and Dalton, teaching them how to ride bike, the list could go on, and to think that the chance for a normal life wouldn't be possible without the dietary treatment is quite heartbreaking. Thank you very much for allowing us to speak to you today, and thank you for your consideration of this matter.

Thankfully!



David Aakre

Mr. Chairman and members of the committee

My life with PKU

My name is Danielle Aakre and I have PKU. PKU is something I can enjoy, but somedays when I'm really busy I get frustrated. But then I'm happy that they were able to find a treatment so you don't become mentally or physically handicapped. If I don't do my part, and if I wasn't on my diet I wouldn't be in the school I'm in, and would be mentally handicapped and that would stop me from doing many different things.

Being that I'm on my diet I can go swimming, play basketball, go to my school, being able to enjoy myself, being in volleyball, rollerblading, going to many different field trips, and being comfortable being myself. What is important about having PKU and if I wasn't on the diet I really wouldn't have a life, and would be mentally or physically handicapped.

I help my Dad out with writing up menus and bringing them back and forth to school, and setting an example for my brothers is very important. I hope they'll grow up and not have a problem with PKU. I try to tell them if people ever try to make fun of them tell them everyones different, no ones the same.

I'm able to have more phe. than some kids with PKU so when I go to parties I'll eat a regular piece of pizza without the cheese and meat, and 2" x 2" piece of cake without ice cream. When I go I have to make sure my diet sheet is filled out and that I drink all my formula, and mix it up for the next day.

When I drink my formula it tastes pretty good like some sort of cherry-fruit punch kind of kool-aid. It took a long time to get it to taste OK, and to get used to drinking it. Sometimes it would make me throw - up when I drank it until I got used to it.

I drink 32 oz myself and the brothers drink 16 oz each. I used to drink 24 oz but they changed it to 32 oz so I get more of the nutrition from the formula that helps me grow.

Thank you for letting us come to talk to you about PKU.

Dr. Kathy Wood

Testimony on SB 2374
before the
House Human Services Committee
March 8, 1999

Chairwoman Price, members of the Committee: Good Morning! My name is Dr. Kathy Wood. I am a physician from Grand Forks here to present testimony on Senate Bill 2374. As a physician, I am prepared to address any questions, which you might have concerning the biochemistry, genetics, detection and treatment of PKU. But even more importantly, I am here on behalf of a very special young lady who was diagnosed with PKU 17 years ago—my youngest daughter, Katie.

As a first year medical student at UND back in 1971, I vividly remember traveling to the Grafton State School one day, and seeing a patient who could not walk, talk, or care for herself in any way. I remember thinking how very sad it was that this young woman (who was my age) could have been "normal" had the screening program been in place for PKU only 12 years earlier, when she was born.

Those of you on this committee who are parents or grandparents can probably understand, then, what it must have felt like for me, 10 years after that visit, to hear that my own beautiful baby girl had the very same genetic disorder.

The good news is that my daughter, who is now a junior in high school and a very committed girl's hockey player, is an "A" student who is planning on attending

UND in hopes of being accepted into Medical School 5 years from now.

But this "success" has not come about easily, or without a great deal of sacrifice. Since infancy, Katie has had to drink a synthetic formula on a daily basis. This metabolic food is expensive and, to most of the rest of us, unpalatable. For her, and for all children with PKU, however, it is crucial for normal growth and development, supplying essential amino acids. Katie is a vegetarian, not allowed to eat high protein foods such as meat, fish, poultry, eggs, milk, cheese, ice cream, nuts or flour products. The fruits, vegetable and grains which she is permitted to eat must all be carefully measured, as these, too, impact on her blood phenylalanine level. She must draw her own blood frequently to insure that this level does not get too high, and then titrate the amount and types of foods she eats accordingly. Her growth and development have been monitored continuously since birth, and she regularly receives assistance with her diet from a nutritionist familiar with this disorder.

The National Collaborative Study on PKU has determined that dietary maintenance for children and young adults with PKU is life-long. For our children, there will be no reprieve. They will be dependent on the metabolic foods and food products for the rest of their lives.

When my daughter was born, it was commonly believed that children could be taken off their diets at age 6. It has subsequently been documented, however, that children who did so suffered drops in their IQ's (sometimes as much as 20-30 points), developed learning disabilities, behavioral problems (e.g. hyperactivity, irritability),

neurologic problems and personality disorders (e.g. schizophrenia, depression, panic attacks). Re-institution of the diet for those initially taken off their formulas and low-protein foods has been shown to improve IQ levels and decrease behavioral and mental health problems.

Another issue which arose as young PKU women began having babies is referred to as "Maternal PKU". It was discovered that, unless the mother was strictly maintained on her diet pre-conception and all through her pregnancy, her babies would universally be born retarded, with small heads (microcephaly), congenital heart defects, craniofacial abnormalities, etc., all due to a direct toxic effect on the fetus by the mother's uncontrolled phenylalanine levels.

As a group, we are all aware of, and empathize with, the concerns which you and members of the insurance industry have about insurance mandates. But what are the alternatives, and, for those of you concerned with fiscal impacts, what, indeed, is the most cost-effective approach to take?

When considering costs to the insurance companies of claims for metabolic foods and food products (which, by the way, are subject to deductible and co-insurance withholds), you must also consider what the financial impact to the state of North Dakota could be if these essential dietary products are not made available to those in need. For those children and young adults with PKU who are untreated or not adequately maintained on their diets, as well as the children of untreated PKU mothers, North Dakota could be faced, in a "best-case" scenario, with the costs of special education and mental health treatment

programs. In a "worst-case" scenario, consider the impact of long-term institutional or custodial care, as in the days prior to the screening program mandates. In addition to these costs, the state would also lose future income from the tax-paying status which these children can achieve if maintained on their diets. We believe that any costs incurred by our insurance carriers are quickly redeemable when considering these alternatives.

In summary, I would like to thank you, Chairwoman Price, and members of this committee, for your time and attention. I implore you, on behalf of my daughter, Katie, and the rest of the PKU children and young adults of this state, to issue a "do-pass" recommendation for Senate Bill 2374. I can assure you that, by doing so, you will leave this legislative session with the knowledge that you have done what is right for these young citizens who must live and deal with this disorder for the rest of their lives. Please remember: they are counting on you for their health and for their futures. Thank you!

GRAND FORKS PHD
PUBLIC HEALTH DEPARTMENT
122 South 5th Street, Suite 210 • Grand Forks, ND 58201 • Telephone (701) 746-2525 • Fax (701) 746-2534

2/10/99

Dear Legislators:

I am a licensed, registered dietitian writing in support of Senate Bill # 2374. The purpose of the bill is to require insurance companies to cover the costs of metabolic foods and formula for people with the diseases PKU and MSUD.

Are you familiar with the Food Guide Pyramid? It is a nutrition guideline for the general population. Attached is a copy for you, but I modified it to be the **Food Guide Pyramid for PKU**. Children with PKU and MSUD cannot follow the Food Guide Pyramid. It would cause them to be mentally retarded, suffer mental illness, have learning disabilities, and suffer behavioral and social problems. Pretend for a moment that you were placed on this diet. What would you eat today? What would you eat tomorrow? What would you eat the rest of your life?

Children with PKU and MSUD must consume a special metabolic formula every day for the rest of their lives. The formula provides the nutrients needed for normal growth and development. (The offending amino acid has been "snipped" from the protein chain in this formula.) In addition to the formula, the child can be given foods that are low in protein. These foods are limited to fruits, fruit juices, small portions of some vegetables, pure sugar products, fats and special low-protein foods. **The treatment for PKU and MSUD is following a very strict diet.**

I sing the praises of metabolic formula and low-protein food products that have been developed for people with diseases such as PKU and MSUD. Through special manufacturing processes, low-protein flour, breads, cereals, pastas, rice, cookies, crackers, and even chips have been created. Unfortunately, these low-protein products are very expensive, due to high costs of manufacturing them. **This cost is a barrier to diet adherence.**

If insurance companies are concerned about their costs going up, I would encourage them to raise the premiums for people who create their own diseases through lifestyle (for example, people who smoke or abuse drugs and alcohol.) The children with metabolic diseases did nothing to create their own plight, nor did their parents. PKU and MSUD are rare, genetically-dictated diseases.

Insurance companies must keep in mind that the cost of covering the diet is low compared to the medical costs of mental retardation and developmental disabilities. Even more important is the assurance given to these precious children that they can live normal, healthy lives. For that, there is no price tag. Please support Senate Bill 2374.

Sincerely,

Bev Benda-Moe, LRD

Bev Benda-Moe, LRD
Nutrition Supervisor

ENCL

FOOD GUIDE PYRAMID

A Guide to Daily Food Choices

FOR PKU

The Pyramid is an outline of what to eat each day. It's not a rigid prescription, but a general guide that lets you choose a healthful diet that's right for you. The Pyramid calls for eating a variety of foods to get the nutrients you need and at the same time the right amount of calories to maintain a healthy weight.

Pure sugar + fat OK

Fats, Oils & Sweets
USE SPARINGLY

KEY

These symbols show fats, oils, and added sugars in foods.

- Fat (naturally occurring and added)
- Sugars (added)

NO

Milk, Yogurt, & Cheese Group
2-3 SERVINGS

Meat, Poultry, Fish, Dry Beans, Eggs, & Nuts Group
2-3 SERVINGS

NO

LIMIT

Vegetable Group
3-5 SERVINGS

Fruit Group
2-4 SERVINGS

OK

Bread, Cereal, Rice, & Pasta Group
6-11 SERVINGS

MUST BE LOW PRO

The **Food Guide Pyramid** emphasizes foods from the five food groups shown in the three lower sections of the Pyramid.

Each of these food groups provides some, but not all, of the nutrients you need. Foods in one group can't replace those in another. No one food group is more important than another—for good health, you need them all.

17-114

Source: U.S. DEPARTMENT OF AGRICULTURE and the U.S. DEPARTMENT OF HEALTH AND HUMAN SERVICES.

Provided by: the Education Department of the NATIONAL LIVE STOCK AND MEAT BOARD.