MICROFILM DIVIDER

OMB/RECORDS MANAGEMENT DIVISION
SFN 2053 (2/85) 5M

GREAT SEAL
STATE OF NORTH DAKOTA

ROLL NUMBER

DESCRIPTION

2239

2001 SENATE HUMAN SERVICES

SB 2239

2001 SENATE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB 2239

Senate Human Services Committee

☐ Conference Committee

Hearing Date January 29,2001

Tape Number	Side A	Side B	Meter #
1	X		15.2
1		X	'17.6
Committee Clerk Signature	Carol Kolodziejchuk		

Minutes:

The hearing was opened on SB 2239.

REPRESENTATIVE KLINSKI introduced the bill. The past bill has been an insurance mandate which is a $3,000 cap on metabolic formula and food for children with PKU. Because of a technical error, the cap only applied to the food portion of it, but was unlimited for the formula. The intent was that the cap would cover all areas of metabolic food and formula. The other portion deals with funding of the formula through the Department of Health. There was a section that stated it was based on the family's ability to pay. We found it detrimental to the families and the Health Department should be covering.

SENATOR FLAKOLL sponsor of the bill, spoke in support of the bill. Used pencil demonstration to illustrate how the body can or cannot break down protein.

SENATOR FISCHER, sponsor of the bill spoke in support of bill. Acknowledged the involvement of many people. Amendments were offered. This would provide an appropriate the funds necessary to cover food and formula as families need it.

DR. TODD TWOGOOD, pediatrician at Medcenter One, supports bill with written testimony.

JACK MCDONALD, ARC of ND, supports bill (Written testimony) SENATOR POLOVITZ: Is this amendment not to have the insurance companies involved with this at all? MR. MCDONALD: Not quite right. The amendments will mean that the Health Department will fund the formula provision from Health Department funds. It would not seek reimbursement from the children's insurance policy. Therefore the full amount of the insurance would be available to the parents for the low-protein food that the health department does not provide. The insurance companies would be involved, but through the parents of these children.

ROD ST. AUBYN, BCBS, supports bill and amendments. (Written testimony)

KAREN EHRENS, ND Dietetic Association, supports bill (Written testimony)

BRENDA RAKOCZY, PKU family, supports bill in written testimony.

DARLENE BARTZ, Department of Health, supports bill, (Written testimony)

ANNE CHRISTIANS, PKU adult, supports bill (Written testimony)

AMY JOHNSON, parent of PKU son, supports this bill, written testimony.

DAVID AAKRE, parent of PKU children, supports this bill in written testimony.

DANIELLE AAKRE, PKU patient, gave speech on PKU.

DAVE PESKE, ND Medical Association, was involved in the discussions; supports bill.

NANCY OUDERKIRK, parent of PKU son, supports bill, written testimony.

Written testimony from KIM and BRENDA ANKENBAUER, parents of PKU children.

RENEE CHIRSTIANS, mother of two young PKU afflicted adults. (Written testimony)

The hearing was closed on SB 2239.

Discussion was resumed on SB 2239, Tape 1, Side B, Meter 17.4.

SENATOR MATHER moved to adopt the amendments offered by Senator Fischer.

SENATOR POLOVITZ seconded the motion. Roll call vote carried 6-0. SENATOR POLOVITZ moved a DO PASS AS AMENDED and rereferred to appropriations. SENATOR FISCHER seconded it. Roll call vote carried 6-0. SENATOR MATHERN will carry the bill.

FISCAL NOTE

Requested by Legislative Council
04/02/2001

Bill/Resolution No.:

Amendment to: Reengrossed
SB 2239

1A. **State fiscal effect:** *Identify the state fiscal effect and the fiscal effect on agency appropriations compared to funding levels and appropriations anticipated under current law.*

	1999-2001 Biennium		2001-2003 Biennium		2003-2005 Biennium	
	General Fund	Other Funds	General Fund	Other Funds	General Fund	Other Funds
Revenues				$140,192		$140,192
Expenditures			$89,152	$140,192	$89,152	$140,192
Appropriations			$89,152	$140,192	$89,152	$140,192

1B. **County, city, and school district fiscal effect:** *Identify the fiscal effect on the appropriate political subdivision.*

1999-2001 Biennium			2001-2003 Biennium			2003-2005 Biennium		
Counties	Cities	School Districts	Counties	Cities	School Districts	Counties	Cities	School Districts

2. **Narrative:** *Identify the aspects of the measure which cause fiscal impact and include any comments relevant to your analysis.*

This bill requires the Department of Human Services to provide medical food or formula and low-protein modified food products to individuals with phenylketonuria (PKU) or maple syrup urine disease (MSUD). It is estimated that 27 individuals will utilize this program during the 2001-2003 biennium. This bill transfers the process of purchasing and distributing formula and low-protein modified food products from the Health Department to the Department of Human Services.

3. **State fiscal effect detail:** *For information shown under state fiscal effect in 1A, please:*
   A. **Revenues:** *Explain the revenue amounts. Provide detail, when appropriate, for each revenue type and fund affected and any amounts included in the executive budget.*

The revenues are anticipated to be derived from the federal Maternal & Child Health Services Block Grant, and from individuals who purchase medical food from the Department at cost.

   B. **Expenditures:** *Explain the expenditure amounts. Provide detail, when appropriate, for each agency, line item, and fund affected and the number of FTE positions affected.*

The expenditures reflected above are based upon purchasing and shipping medical food and low-protein modified food products for an estimated 27 individuals.

   C. **Appropriations:** *Explain the appropriation amounts. Provide detail, when appropriate, of the effect* *on the biennial appropriation for each agency and fund affected and any amounts included in the executive budget. Indicate the relationship between the amounts shown for expenditures and appropriations.*

The Department of Human Services appropriation would need to be increased by $229,344, with $89,152 to purchase the necessary medical foods for program participants, as provided in this bill.

If the Department's appropriation is not increased to cover this additional program, the service contracts for multidisciplinary clinics, care coordination and family support activities would have to be reduced.

Name:	Debra A. McDermott	**Agency:**	Department of Human Services
Phone Number:	328-3695	**Date Prepared:**	04/04/2001

FISCAL NOTE

Requested by Legislative Council

2/20/2001

Bill/Resolution No.:

Amendment to: Engrossed
SB 2239

1A. **State fiscal effect:** *Identify the state fiscal effect and the fiscal effect on agency appropriations compared to funding levels and appropriations anticipated under current law.*

	1999-2001 Biennium		2001-2003 Biennium		2003-2005 Biennium	
	General Fund	Other Funds	General Fund	Other Funds	General Fund	Other Funds
Revenues				$80,000		$80,000
Expenditures				$80,000		$80,000
Appropriations						

1B. **County, city, and school district fiscal effect:** *Identify the fiscal effect on the appropriate political subdivision.*

1999-2001 Biennium			2001-2003 Biennium			2003-2005 Biennium		
Counties	Cities	School Districts	Counties	Cities	School Districts	Counties	Cities	School Districts

2. **Narrative:** *Identify the aspects of the measure which cause fiscal impact and include any comments relevant to your analysis.*

Due to increased costs of metabolic food (formula) provided to individuals with phenylketonuria (PKU) and maple syrup urine disease (MSUD) and the increased progam participation, it is estimated that an increase of $80,000 will be incurred. The reengrossed bill allows the Department to seek reimbursment from any government program that provides coverage to that individual. For the 01-03 biennium it is estimated that we may collect approximately $8,000 from other government programs. **Included in the Health Department's appropriation bill SB 2004 is $80,000 from the community health trust fund.**

3. **State fiscal effect detail:** *For information shown under state fiscal effect in 1A, please:*
   A. **Revenues:** *Explain the revenue amounts. Provide detail, when appropriate, for each revenue type and fund affected and any amounts included in the executive budget.*

The $80,000 in the revenue column is from the community health trust fund and is included in the Health Department's appropriation bill SB 2004

   B. **Expenditures:** *Explain the expenditure amounts. Provide detail, when appropriate, for each agency, line item, and fund affected and the number of FTE positions affected.*

It is estimated that the increased costs of medical foods and the increased program participation would cost an additional $80,000 for the 2001-03 and 2003-05 bienniums. It is included in the Health Department's appropriation bill SB 2004.

C. **Appropriations:** *Explain the appropriation amounts. Provide detail, when appropriate, of the effect on the biennial appropriation for each agency and fund affected and any amounts included in the executive budget. Indicate the relationship between the amounts shown for expenditures and appropriations.*

Name:	Kathy J. Albin	**Agency:**	Department of Health
Phone Number:	328-2392	**Date Prepared:**	03/05/2001

FISCAL NOTE

Requested by Legislative Council

02/01/2001

Bill/Resolution No.:

Amendment to: SB 2239

1A. **State fiscal effect:** *Identify the state fiscal effect and the fiscal effect on agency appropriations compared to funding levels and appropriations anticipated under current law.*

	1999-2001 Biennium		2001-2003 Biennium		2003-2005 Biennium	
	General Fund	Other Funds	General Fund	Other Funds	General Fund	Other Funds
Revenues						
Expenditures			$80,000		$80,000	
Appropriations						

1B. **County, city, and school district fiscal effect:** *Identify the fiscal effect on the appropriate political subdivision.*

1999-2001 Biennium			2001-2003 Biennium			2003-2005 Biennium		
Counties	Cities	School Districts	Counties	Cities	School Districts	Counties	Cities	School Districts

2. **Narrative:** *Identify the aspects of the measure which cause fiscal impact and include any comments relevant to your analysis.*

Due to the increased costs of metabolic food (formula) provided to individuals with phenylketonuria (PKU) and maple syrup urine disease (MSUD), and the increased program participation, it is estimated that an increase of $80,000 of general funds will be incurred.

3. **State fiscal effect detail:** *For information shown under state fiscal effect in 1A, please:*
   A. **Revenues:** *Explain the revenue amounts. Provide detail, when appropriate, for each revenue type and fund affected and any amounts included in the executive budget.*

   B. **Expenditures:** *Explain the expenditure amounts. Provide detail, when appropriate, for each agency, line item, and fund affected and the number of FTE positions affected.*

It is estimated that increased costs of medical foods and the increased program participation would cost an additional $80,000 for the 2001-03 and the 2003-05 bienniums.

   C. **Appropriations:** *Explain the appropriation amounts. Provide detail, when appropriate, of the effect on the biennial appropriation for each agency and fund affected and any amounts included in the executive budget. Indicate the relationship between the amounts shown for expenditures and appropriations.*

If this bill is passed the Department will need $80,000 of increased general fund appropriation. These costs are not included in our 2001-03 budget request SB 2004.

Name:	Kathy J. Albin	**Agency:**	Health Department
Phone Number:	328-2392	**Date Prepared:**	02/01/2001

FISCAL NOTE

Requested by Legislative Council
01/17/2001

Bill/Resolution No.: SB 2239

Amendment to:

1A. **State fiscal effect:** *Identify the state fiscal effect and the fiscal effect on agency appropriations compared to funding levels and appropriations anticipated under current law.*

	1999-2001 Biennium		2001-2003 Biennium		2003-2005 Biennium	
	General Fund	Other Funds	General Fund	Other Funds	General Fund	Other Funds
Revenues				$70,000		$70,000
Expenditures			$10,000	$70,000	$10,000	$70,000
Appropriations						

1B. **County, city, and school district fiscal effect:** *Identify the fiscal effect on the appropriate political subdivision.*

1999-2001 Biennium			2001-2003 Biennium			2003-2005 Biennium		
Counties	Cities	School Districts	Counties	Cities	School Districts	Counties	Cities	School Districts

2. **Narrative:** *Identify the aspects of the measure which cause fiscal impact and include any comments relevant to your analysis.*

Due to the increased costs of metabolic food (formula) provided to individuals with phenylketonuria (PKU) and maple syrup urine disease (MSUD), and the increased program participation, it is estimated that an increase of $80,000 will be incurred. It is anticipated that $70,000 will be collected from insurance companies, nonprofit health service corporations, health maintenance organizations, and government programs. The remaining $10,000 will be needed from the general fund.

3. **State fiscal effect detail:** *For information shown under state fiscal effect in 1A, please:*
   A. **Revenues:** *Explain the revenue amounts. Provide detail, when appropriate, for each revenue type and fund affected and any amounts included in the executive budget.*

It is estimated that the Department will collect $70,000 from insurance companies, nonprofit health service corporations, health maintenance organizations, and government programs.

   B. **Expenditures:** *Explain the expenditure amounts. Provide detail, when appropriate, for each agency, line item, and fund affected and the number of FTE positions affected.*

It is estimated that increased costs of medical foods and the increased program participation would cost an additional $80,000 for the 2001-03 biennium. It is anticipated that of the $80,000 of additional expenses that $70,000 will be reimbursed by some from of insurance provider or government program and that $10,000 of general funds will be needed.

C. **Appropriations:** *Explain the appropriation amounts. Provide detail, when appropriate, of the effect on the biennial appropriation for each agency and fund affected and any amounts included in the executive budget. Indicate the relationship between the amounts shown for expenditures and appropriations.*

If this bill is passed the Department will need $80,000 of increased appropriation. These costs are not included in our appropriation SB 2004.

Name:	Kathy J. Albin	**Agency:**	Health
Phone Number:	328-2392	**Date Prepared:**	01/23/2001

Senator Tom Fischer

Proposed amendments to SB 2239

Page 3, line 10 after income. delete "If treatment services under this subsection are

Page 3 delete lines 11 thru 14

Page 3 line 19 delete "3 and

Page 5 remove the overstrike on section 4. lines 11 thru 13

Page 5 line 13 delete "Any insurance

Page 5 delete lines 14 thru 17

Date: 1/29/01

Roll Call Vote #: 1

2001 SENATE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. 2239

Senate HUMAN SERVICES Committee

☐ Subcommittee on

or

☐ Conference Committee

Legislative Council Amendment Number

Action Taken Amendment

Motion Made By Sen Mathern Seconded By Sen Polovitz

Senators	Yes	No	Senators	Yes	No
Senator Lee, Chairperson	✓		Senator Polovitz	✓	
Senator Kilzer, Vice-Chairperson	✓		Senator Mathern	✓	
Senator Erbele	✓				
Senator Fischer	✓				

Total (Yes) 6 No 0

Absent 0

Floor Assignment

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

Sen Fischer's amendments

Date: 1/29/01

Roll Call Vote #: 2

2001 SENATE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. 2239

Senate HUMAN SERVICES Committee

☐ Subcommittee on

or

☐ Conference Committee

Legislative Council Amendment Number

Action Taken Do Pass

Motion Made By Sen Polovitz Seconded By Sen Fischer

Senators	Yes	No	Senators	Yes	No
Senator Lee, Chairperson	✓		Senator Polovitz	✓	
Senator Kilzer, Vice-Chairperson	✓		Senator Mathern	✓	
Senator Erbele	✓				
Senator Fischer	✓				

Total (Yes) 6 No 0

Absent 0

Floor Assignment Sen Mathern

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

Module No: SR-16-1928
Carrier: T. Mathern
Insert LC: 10100.0401 Title: .0500

REPORT OF STANDING COMMITTEE

SB 2239: Human Services Committee (Sen. Lee, Chairman) recommends **AMENDMENTS AS FOLLOWS** and when so amended, recommends **DO PASS** and **BE REREFERRED** to the **Appropriations Committee** (6 YEAS, 0 NAYS, 0 ABSENT AND NOT VOTING). SB 2239 was placed on the Sixth order on the calendar.

Page 3, line 10, remove "If treatment services under this subsection are"

Page 3, remove lines 11 through 14

Page 3, line 19, replace "subsections" with "subsection" and remove "3 and"

Page 5, line 11, remove the overstrike over "~~This section does not require medical benefits coverage for low protein modified~~"

Page 5, remove the overstrike over line 12

Page 5, line 13, remove the overstrike over "~~available to that individual under a department of health program.~~" and remove "Any insurance"

Page 5, remove lines 14 through 17

Renumber accordingly

2001 SENATE APPROPRIATIONS

SB 2239

2001 SENATE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB 2239

Senate Appropriations Committee

☐ Conference Committee

Hearing Date February 9, 2001

Tape Number	Side A	Side B	Meter #
1	X		39.1-54.6
		X	0.0-15.6
Committee Clerk Signature			

Minutes:

Senator Solberg opened the hearing on SB 2239.

Darleen Bartz, Chief of the Preventive Health Section of the State Department Health, (testimony attached and Chart).

Senator Solberg: Explain the two fiscal notes?

Darlene Bartz: Amendments were done; if this bill is passed, $80,000 will come out of the general fund due to increased costs of the medical food provided to individuals with PKU and MSUD and the increased program participation.

Senator Andrist: How much would the department have spent with the same rules as stated here; $80,000 is not really a reliable figure?

Darlene Bartz: It is with projections and changes; diet and formula go into this consideration; there was no new bonus in the last three years; new formulas have now been developed.

Senator Judith Lee, District 13, spoke in favor of the bill and asked the committee to give this serious consideration. The amendment was needed because it was unfair to the families with insurance. The child disease PKU has been prevented by children taking these food products. Past stories has helped with this program.

Senator Tom Fisher, District 46, spoke in support of the bill. It is important that people receive these benefits; return in the investment is substantial.

Representative Amy Kliniske, District 42 spoke in favor of the bill. This program helps after insurance to stay on the diet for life, mainly PKU. The total cap per child is $3,000.00 for diet and food which they have to stay on for life. Formula was left unlimited with past bills to meet block grant. Fiscal note is base don three born next biennium; projects 1-10,000 born with PKU

End Tape #1, Side A, meter 54.6

Start Tape #1, Side B, meter 0.0

And the three births is realistic. The new formula costs double from the formula the children receive now. It tastes better as the old formula was sulfur based. The families that chose the new formula will pay the extra cost or the difference. The formula is purchased from the Health Department at costs within the certain age limit.

Senator Solberg: This is a rule making process; expand on how close this bill relates to rules that were rejected by the rules committee?

Amy Kliniske: It was a direct result. The rule making on paragraph 2 states department is to provide based on the families ability to pay. Standard rule making process, income/insurance 150% poverty first rule, 2nd rule 185% of poverty considering cost. Promises have been made to families on this rule making. There are difficult needs for different kids with the food and formula for cost differences. High calories and fat in the formula. The kids eat very little considering their weight.

Tape #1, Side B, meter 8.3

Rod St. Aubyn, spoke representing Blue Cross/Blue Shield of North Dakota (testimony attached) which explained insurance view of the program. Asked for support of the bill and the appropriation.

Darlene Bartz: Commented. There are two projects births next biennium. Concerns which were amended to collect Medicaid and wick which was taken out. The reason was the general funds language and this was eliminated by the amendment.

Senator Heitkamp: What's the turn back; where do we go?

Darlene Bartz: $80,000 line item; it was not used and turned back; the past this was out of MCH Block Grant; Different percentage $120,000 block grant with general funds and general funds. Additional $80,000 for costs with the program.

Dave Peske, ND Medical Association, stated all physicians in North Dakota/pediatrics favor and support this bill.

Senator Andrist: Suggest we have amendments prepared to the effect to recapture moneys without additional responsibility to Medicaid/Medicare.

Senator Solberg: This will be looked at.

With no opposition, the hearing was closed. Tape #1, Side B, meter 15.6

February 13, 2001 Full Committee Action (Tape #3, Side A; Meter No. 12-0-17.8)

Senator Nething reopened the testimony on SB2239.

Senator Andrist presented amendment #10100.0501. Discussion on the amendment.

Senator Andrist moved the amendment be adopted; seconded by Senator Holmberg. Verbal discussion approved motion.

Discussion on the bill.

Senator Nething noted the amendment adopted; but the bill should be sent back to the Health Subcommittee for further review.

February 16, 2001 Full Committee Action (Tape #2, Side A; Meter No. 1.2-3.4)

Senator Nething reopened the hearing on SB2239.

Senator Andrist, Chair of the Health Subcommittee, moved a DO PASS AS AMENDED ; Senator Holmberg seconded. The amendment had been adopted on 2-13-01. Roll Call Votes: 14 yes; 0 no, 0 absent and not voting

10100.0501
Title. .0600

Prepared by the Legislative Council staff for
Senator Andrist
February 12, 2001

PROPOSED AMENDMENTS TO ENGROSSED SENATE BILL NO. 2239

Page 3, line 10, after the underscored period insert "If treatment services under this subsection are provided to an individual by the department, the department may seek reimbursement from any government program that provides coverage to that individual for the treatment services provided by the department."

Page 3, line 15, replace "subsection" with "subsections 3 and"

Renumber accordingly

16

Date: 2-13-01

Roll Call Vote #:

2001 SENATE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. SB 2239

Senate Appropriations Committee

☐ Subcommittee on

or

☐ Conference Committee

Legislative Council Amendment Number 10100.0501 (adopt 2-13)

Action Taken Do pass as amended

Motion Made By Senator Andrist Seconded By Senator Holmberg

Senators	Yes	No	Senators	Yes	No
Dave Nething, Chairman	✓				
Ken Solberg, Vice-Chairman	✓				
Randy A. Schobinger	✓				
Elroy N. Lindaas	✓				
Harvey Tallackson	✓				
Larry J. Robinson	✓				
Steven W. Tomac	✓				
Joel C. Heitkamp	✓				
Tony Grindberg	✓				
Russell T. Thane	✓				
Ed Kringstad	✓				
Ray Holmberg	✓				
Bill Bowman	✓				
John M. Andrist	✓				

Total Yes 14 No 0

Absent 0

Floor Assignment Senator Andrist

If the vote is on an amendment, briefly indicate intent: (Camera / Sen Mathern bill)

REPORT OF STANDING COMMITTEE (410)
February 16, 2001 3:56 p.m.

Module No: SR-29-3780
Carrier: Andrist
Insert LC: 10100.0501 Title: .0600

REPORT OF STANDING COMMITTEE

SB 2239, as engrossed: Appropriations Committee (Sen. Nething, Chairman) recommends **AMENDMENTS AS FOLLOWS** and when so amended, recommends **DO PASS** (14 YEAS, 0 NAYS, 0 ABSENT AND NOT VOTING). Engrossed SB 2239 was placed on the Sixth order on the calendar.

Page 3, line 10, after the underscored period insert "If treatment services under this subsection are provided to an individual by the department, the department may seek reimbursement from any government program that provides coverage to that individual for the treatment services provided by the department."

Page 3, line 15, replace "subsection" with "subsections 3 and"

Renumber accordingly

2001 HOUSE HUMAN SERVICES

SB 2239

2001 HOUSE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB 2239

House Human Services Committee

☐ Conference Committee

Hearing Date March 12, 2001

Tape Number	Side A	Side B	Meter #
1	x		1 to 3362
3	x		1100 to 1550
Committee Clerk Signature Corinne Caston			

Minutes:

Vice Chairman Devlin: I will call the committee to order. Clerk call the roll. We will open the hearing on SB 2239.

Sen. Tom Fischer - District 46: I am here to introduce and ask your support of SB 2239 dealing with PKU. This has been a work in progress, but I think we've got it right. The only amendment that was made in appropriations has to do with reimbursement from any government program and where those dollars are to be deposited. I would ask for your support of SB 2239.

Rep. Kliniske - District 42: This is an insurance mandate that required insurance companies to cover the medical foods, which is the formula and foods portion for children who are PKU. Basically that mandate was intended to be capped at $3000. The first session that mandate passed with the cap and a sunset clause. Automatically we had to bring it back the next session because of the sunset clause to make it permanent. The effect on insurance was a total of $18,000 at that time, yet it was a huge benefit for families that needed that coverage. When we came back last session, we passed the bill as it was and after we left, we realized there were some glitches in the bill. Basically 2 problems we found. The first problem derived from the Attorney Generals opinion that stated that the $3000 cap only applies to the food portion, but the formula portion was left wide open. We had never intended that. The 2nd portion of the bill dealt with language that was in the original bill from 1967. In that bill were three paragraphs that described the program. Within the second paragraph, the last sentence said, "based on families ability to pay". Those words, once they were discovered in the health department, required the health department to start a rule making process to determine what a families ability to pay was. We as sponsors understood so well, that that was a responsibility that the health department had to take on. However, during the process, the limits that were established were not acceptable to the families. The health department decided that we should take care of this process at the next Legislative Session and that is why we are here today. That language will be removed from the bill. Basically it will say that the health department will cover these children. That is the bill before you.

Brenda Rakoczy - Mother of PKU Child: (See written testimony).

Rep. Galvin: In your one paragraph you referred to the ability to purchase these materials at cost from the health department and the astronomical mark up in the pharmacy. How astronomical are these mark ups?

Rakoczy: May I refer that to Mrs. Christians.

Vice Chair Devlin: Any further testimony in support of SB 2239?

Renee Christians: I am here today in support of SB 2239 (See written testimony).

Ann Christian: I am here today in support of SB 2239. (See written testimony).

Jack McDonald - Rep. Of the ARC of ND: We strongly support this bill. (See written testimony).

Dr. Bob Wentz - Pediatrician and a State Health Officer: I am here in support of SB 2239 (See written testimony). (Also entered into testimony by Dr. Wentz was testimony from Dr. Todd Twogood.)

Darleen Bartz - Chief of the Preventative Health Section of the Dept. Of Health: (See written testimony).

Rep. Metcalf: Currently how many people are being served by Department?

Bartz: 18.

Rep. Weisz: Has that number been static over the years?

Bartz: It has kinda changed over the years, we are seeing an increase of people who are coming back on to this program. We have not had new births for approximately 3 years and usually we are seeing a new birth one every two years. So we are anticipating that.

David Peske - ND Medical Association: You've heard from three prior witnesses that both the Medical Association and the Academy of Pediatrics were involved in developing this bill and were in full support of that, and I wanted to confirm that.

Rep. Kliniske: Blue Cross/Blue Shield are in support of this bill and you have a copy of their written testimony. (See written testimony).

Vice Chair Devlin: Any further testimony in support of SB 2239? Any opposition? I will close the hearing on SB 2239.

COMMITTEE WORK:

CHAIRMAN PRICE: Let's go to SB 2239. You all have the fiscal note dated 2/20? It says "Included in the Health Department' appropriation bill, SB 2004 is $80,000 from the Community Health Trust Fund". There is nothing in the bill here that says where the money is coming from.

Personally I don't want to start funding other programs out of the Tobacco Fund. My intention is that when this goes through Appropriations, that we tell them that is not our desire. It should just be General Fund dollars. I got something from the Legislative Council that says there are four things they are trying to fund out of Community Trust Fund, and this is one of them. The other was the Dental Program and two other things.

REP. GALVIN: Move a DO PASS.

REP. TIEMAN: Second.

CHAIRMAN PRICE: Any other discussion? The clerk will take the roll on a **DO PASS and be rereferred to Appropriations.**

14 YES 0 NO 0 ABSENT REP. GALVIN

Date: 3-12-01
Roll Call Vote #: 1

2001 HOUSE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. SB 2239

House Human Services Committee

☐ Subcommittee on

or

☐ Conference Committee

Legislative Council Amendment Number

Action Taken DO PASS & rerefer to Appropriations

Motion Made By Rep. Galvin Seconded By Rep. Tieman

Representatives	Yes	No	Representatives	Yes	No
Clara Sue Price - Chairman	✓		Audrey Cleary	✓	
William Devlin - V. Chairman	✓		Ralph Metcalf	✓	
Mark Dosch	✓		Carol Niemeier	✓	
Pat Galvin	✓		Sally Sandvig	✓	
Frank Klein	✓				
Chet Pollert	✓				
Todd Porter	✓				
Wayne Tieman	✓				
Dave Weiler	✓				
Robin Weisz	✓				

Total (Yes) 14 No 0

Absent 0

Floor Assignment Rep. Galvin

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

REPORT OF STANDING COMMITTEE

SB 2239, as reengrossed: Human Services Committee (Rep. Price, Chairman) recommends **DO PASS** and **BE REREFERRED** to the **Appropriations Committee** (14 YEAS, 0 NAYS, 0 ABSENT AND NOT VOTING). Reengrossed SB 2239 was rereferred to the **Appropriations Committee.**

2001 HOUSE APPROPRIATIONS

SB 2239

2001 HOUSE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB2239

House Appropriations Committee

☐ Conference Committee

Hearing Date **March 20, 2001**

Tape Number	Side A	Side B	Meter #
1		x	5779 - 6212
2	X		0 - 1251
Committee Clerk Signature			

Minutes:

HOUSE APPROPRIATIONS COMMITTEE HEARING ON SB2239.

Rep. Timm: Opened the hearing on SB2239.

Rep. Kliniske: Many of you may remember that we have dealt with the PKU issue in the past and order to get you through this bill I need to give you just a brief history of why were here today. The first bill back in 1997 established an insurance mandate to cover PKU metabolic food and low protein modified food and a cap of $3000 for children with the disease. There was a sunset clause placed on that to see what the effect would be in the next biennium to see if we could continue that, when we came back in the next biennium we found out that only six families had the appropriate coverage, therefore the maximum that would be charged out to insurance companies was $18,000 and of that $18,000 only $12,000 has actually been claimed on insurance and so those families that did have the insurance coverage were utilizing it. What the bill in 1999 was supposed to do was to remove the sunset clause, and shortly after we left in 1999 we found a couple of technical errors with the bill. Number one, in the Attorney General's opinion, she reviewed the bill and determined that the cap applied only to the low protein modified food portion but did not apply to the formula therefore we left the formula wide open when sponsors had intended to place that cap, and that's one of the main reasons we are back here right now is the integrity of the sponsors and the integrity of the families was at stake, and we always intended that it would be a $3000 cap total not a cap just on the food and not on the formula and so this bill will clarify that the cap applies to both The second change in the bill, the original bill that mandated the screening for PKU was back in 1967. In that original bill there was in paragraph two a section that talked about the health department funding the formula for these families based on the families ability to pay. After we left in the 1999 session the health department and rightly so, determined that they needed to go through a rule making process to determine what that meant on the families ability to pay, unfortunately what happened during the rule making process was objectionable both to the sponsor and to the families and pretty much the entire medical community and so the health council suspended the rule making process until we could come here to clarify exactly what that should be, we have as sponsors and families and health professionals determined that it is in the best interest of the State of North Dakota that we continue the practice that has taken place in health department for the past 34 years, and that is that the health department cover the costs of the formula and then the families pick up the cost of the low protein food. Males are eligible for funding up to the age of 22 and females up to the age of 44, so once they hit those caps they are no longer they are no longer eligible for funding through the health department. This bill states that those individuals as opposed to having to go to a pharmacy and paying a %300 markup, those individuals would be able to purchase the formula from the health department at cost once they are no longer eligible. Basically, that outlines the bill and what it does and many of you have been through the PKU issue before and are familiar with what the disease is and how extremely expensive this disease is, I will give you a prime example, in food costs alone and I'm going to use one of the families who has a daughter who is five years old and as she grows older she is going to need more and more of the formula because that is where she gets her nutrition they cannot have dairy products, they cannot have meat, they can't have legumes, no beans and no grain, basically there foods are low protein modified foods, or synthetic foods. This girl is only five years old and here foods costs are already on a monthly basis exceed the families mortgage payment.

Rep. Timm: Any questions of Rep. Kliniske?

Rep. Delzer: In looking through the bill is there any changes in the requirement or the way the testing is done and I see that newborn is added all over and yet wasn't it newborn before?

Rep. Kliniske: I'm not exactly sure if its just a change in where it is written, because this is a new section and so we have to add it to this section.

Rep. Wald: The insurance mandate is in current law right?

Rep. Kliniske: Yes, and it should be noted that if we do not pass this bill the mandate goes back to unlimited formula coverage and limited food coverage, this bill would limit them both to the $3000 cap, so without this bill we go to unlimited.

Rep. Wald: Why did you use regardless of income on page 3 at least twice that I see here?

Rep. Kliniske: That goes back to the second situation that I talked about with the rule making proceedings, what happened is that the original legislation had the words "based on the families ability to pay" and what happened is that the rule that was proposed eliminated all but two families from receiving help from the health department that had been receiving funding for the last 34 years.

Rep. Svedjan: Just for clarification, I'm looking at the health department bill and it does reference in there based on the senate action that there is an appropriation of $80,000, but in your testimony you indicated that funding for the PKU issue comes out of the maternal child health block grant, the $80,000 referenced in here comes from the community health trust fund. Could you clarify that for us?

Rep. Kliniske: The maternal and child health block grant it was a policy decision based on the department that they would use up to $120,000 of that grant or that funding to cover this program, the are asking for the additional what they anticipate to be $80,000 of funding to come from what originally was to be general funds and in the House Human Services was changed to be Community Health Block Grant.

Rep. Timm: Any other questions?

Sen. Fischer: I stand in support of SB2239 better known as the PKU bill and also here doing duty for Sen. Lee who asked me to lend her support to the bill as well as Blue Cross and Blue Shield. As I said in following Rep. Kliniske I don't think there is much that I can add to this other than hopefully that this will be the final time that this will put before the legislator I think we finally have it right and would ask for your support.

Rep. Timm: Any questions?

Jack McDonald, Representing the Arc of North Dakota. (Had written testimony and answered questions following the written testimony that was presented)

Rep. Kempenich: Do this kids have any other medical problems too or is this mainly the one that they have?

Mr. McDonald: This is basically the main problem that they have.

Rep. Kliniske: Most of the children stay on the diet and if they follow there diet then this is there only medical problem. If the children are not on the diet from birth they will become mentally retarded, if they go on the diet for a number of years and then later life if they chose to come off of the diet, but his is a life long diet, if they come off of the diet, they develop problems such as mental problems, disorderly conduct, and all sorts of problems they can't concentrate they become euphoric, its many, many problems associated with the disease.

Rep. Timm: Any more questions for Mr. McDonald?

Darleen Bartz, Department of Health: (Followed written testimony and then answered questions after her testimony)

Rep. Timm: Women up to 45 are covered, are these women encouraged maybe to not have children or doesn't it make any difference?

Ms. Bartz: No it doesn't make any difference.

Rep. Skarphol: When you refer to rule making authority I see it in section three but I'm assuming it does not exempt you in any way from the administrative rule process is that correct?

Ms. Bartz: Yes, that is correct.

Rep. Koppleman: Do you see the rule making authority here as strictly defining what a metabolic disease is or does it go beyond that?

Ms. Bartz: That's what I see as the main piece of it is, as time goes by there are new metabolic diseases that are discovered that there may be a need for testing for and what this would allow is for the Health Council to look at that.

Rep. Timm: Any other questions? Any other testimony in support of SB2239? Any opposition to SB2239? Then we will close the hearing on SB2239.

HOUSE APPROPRIATIONS COMMITTEE HEARING ON SB2239 COMPLETED.

2001 HOUSE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB2239A

House Appropriations Committee

☐ Conference Committee

Hearing Date **March 21, 2001**

Tape Number	Side A	Side B	Meter #
1		x	466 - 699
Committee Clerk Signature			

Minutes:

HOUSE APPROPRIATIONS COMMITTEE ACTIN ON SB2239A.

Rep. Timm: This is the PKU bill is that correct? Moved by Rep. Kliniske for a DO PASS, seconded by Rep. Thoreson. Any discussion?

Rep. Wald: We have been mandating this benefit all along? Response was yes, we have been mandating this since 1997.

Rep. Timm: Any other discussion? We will call the roll for a DO PASS. (21) YES (0) NO. Motion passes. Rep. Kliniske will carry the bill to the floor.

End of House Appropriations Committee action on SB2239A.

2001 HOUSE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB 2239

House Appropriations Committee

☐ Conference Committee

Hearing Date March 28, 2001

Tape Number	Side A	Side B	Meter #
03-28-01 tape #1	0 - 85		
Committee Clerk Signature			

Minutes:

The committee was called to order, and opened committee work on SB 2239.

<u>Chairman Timm</u>: We have the bill in committee. What do we want to do with it?

<u>Rep. Kliniski</u>: She is working on amendments for that right now. Hopes to have the amendments done later today.

The chairman closed the committee work on this bill.

2001 HOUSE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. SB 2239

House Appropriations Committee

☐ Conference Committee

Hearing Date March 30, 2001

Tape Number	Side A	Side B	Meter #
03-30-01 tape #1	1159 - 1649		
Committee Clerk Signature Kathie Hall			

Minutes:

The committee was called to order, and opened committee work on SB 2239.

Rep. Kliniski: Explains proposed amendment. Taking this out of the department of health and putting it into the department of human services in children's special health services, where it rightly belongs.

Rep. Byerly: Moves to reconsider previous action. Seconded by Rep. Delzer.

Voice vote reconsiders action. Motion passes.

Rep. Kliniski: Moves to adopt the amendment. Seconded by Rep. Thoreson.

Voice vote adopts the amendment.

Rep. Koppelman: Moves DO PASS AS AMENDED. Seconded by Rep. Warner.

Vote on Do Pass as Amended : 20 yes, 1 no, 0 absent and not voting.

Rep. Kliniski is assigned to carry this bill to the floor.

Date: 3/21/01
Roll Call Vote #: 1

2001 HOUSE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. SB 2239

House APPROPRIATIONS Committee

☐ Subcommittee on
or
☐ Conference Committee

Legislative Council Amendment Number

Action Taken DO PASS

Motion Made By KLINISKE Seconded By THORESON

Representatives	Yes	No	Representatives	Yes	No
Timm - Chairman	✓				
Wald - Vice Chairman	✓				
Rep - Aarsvold	✓		Rep - Koppelman	✓	
Rep - Boehm	✓		Rep - Martinson	✓	
Rep - Byerly	✓		Rep - Monson	✓	
Rep - Carlisle	✓		Rep - Skarphol	✓	
Rep - Delzer	✓		Rep - Svedjan	✓	
Rep - Glassheim	✓		Rep - Thoreson	✓	
Rep - Gulleson	✓		Rep - Warner	✓	
Rep - Huether	✓		Rep - Wentz	✓	
Rep - Kempenich	✓				
Rep - Kerzman	✓				
Rep - Kliniske	✓				

Total (Yes) 21 No 0

Absent 0

Floor Assignment KLINISKE

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

Date: 3-30-01
Roll Call Vote #: 1

2001 HOUSE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. SB 2239

House APPROPRIATIONS Committee

☐ Subcommittee on
or
☐ Conference Committee

Legislative Council Amendment Number

Action Taken Motion to reconsider previous action

Motion Made By Rep Byerly Seconded By Rep. Delzer

Representatives	Yes	No	Representatives	Yes	No
Timm - Chairman					
Wald - Vice Chairman					
Rep - Aarsvold			Rep - Koppelman		
Rep - Boehm			Rep - Martinson		
Rep - Byerly			Rep - Monson		
Rep - Carlisle			Rep - Skarphol		
Rep - Delzer			Rep - Svedjan		
Rep - Glassheim			Rep - Thoreson		
Rep - Gulleson			Rep - Warner		
Rep - Huether			Rep - Wentz		
Rep - Kempenich					
Rep - Kerzman					
Rep - Kliniske					

Voice vote

Total (Yes) No

Absent

Floor Assignment

passes

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

10100.0601
Title.

Prepared by the Legislative Council staff for
Representative Kliniske
March 28, 2001

PROPOSED AMENDMENTS TO REENGROSSED SENATE BILL NO. 2239

Page 1, line 1, after "25-17" insert "and a new subsection to section 50-10-06"

Page 1, line 2, after "law" insert "and services for treatment of phenylketonuria and maple syrup urine disease"

Page 2, line 24, after "council" insert "and the department of human services"

Page 2, line 29, after the second period insert:

"1."

Page 3, line 1, overstrike "1." and insert immediately thereafter "a."

Page 3, line 4, overstrike "2." and insert immediately thereafter "b."

Page 3, remove lines 8 through 24

Page 3, line 25, replace "7" with "c"

Page 3, after line 26, insert:

"2. The department of human services, as a program provided under chapter 50-10, shall:

a. Provide medical food at no cost to males under age twenty-two and females under age forty-five who are diagnosed with phenylketonuria or maple syrup urine disease, regardless of income. If treatment services under this subsection are provided to an individual by the department, the department may seek reimbursement from any government program that provides coverage to that individual for the treatment services provided by the department.

b. Offer for sale at cost medical food to females age forty-five and over and to males age twenty-two and over who are diagnosed with phenylketonuria or maple syrup urine disease, regardless of income. These individuals are responsible for payment to the department for the cost of medical food.

c. Provide low-protein modified food products, if medically necessary as determined by a qualified health care provider, to females under age forty-five and males under age twenty-two who are receiving medical assistance and are diagnosed with phenylketonuria or maple syrup urine disease."

Page 5, line 12, after "health" insert "or department of human services"

Page 5, after line 12, insert:

"**SECTION 8.** A new subsection to section 50-10-06 of the North Dakota Century Code is created and enacted as follows:

Provide medical food and low-protein modified food products under chapter 25-17 to individuals with phenylketonuria or maple syrup urine disease."

Renumber accordingly

Date: 3-30-01
Roll Call Vote #: 2

2001 HOUSE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. SB2239

House APPROPRIATIONS Committee

☐ Subcommittee on

or

☐ Conference Committee

Legislative Council Amendment Number 10100.0601

Action Taken to adopt amendment

Motion Made By Rep. Kliniske Seconded By Rep. Thoreson

Representatives	Yes	No	Representatives	Yes	No
Timm - Chairman					
Wald - Vice Chairman					
Rep - Aarsvold			Rep - Koppelman		
Rep - Boehm			Rep - Martinson		
Rep - Byerly			Rep - Monson		
Rep - Carlisle			Rep - Skarphol		
Rep - Delzer			Rep - Svedjan		
Rep - Glassheim			Rep - Thoresson		
Rep - Gulleson			Rep - Warner		
Rep - Huether			Rep - Wentz		
Rep - Kempenich					
Rep - Kerzman					
Rep - Kliniske					

Voice Vote

Total (Yes) No

Absent

Floor Assignment

passed

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

Date: 3-30-01
Roll Call Vote #: 3

2001 HOUSE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. SB 2239

House APPROPRIATIONS Committee

☐ Subcommittee on

or

☐ Conference Committee

Legislative Council Amendment Number 10100.0601

Action Taken Do Pass As Amended

Motion Made By Rep. Koppelman Seconded By Rep. Warner

Representatives	Yes	No	Representatives	Yes	No
Timm - Chairman	✓				
Wald - Vice Chairman	✓				
Rep - Aarsvold	✓		Rep - Koppelman	✓	
Rep - Boehm	✓		Rep - Martinson	✓	
Rep - Byerly	✓		Rep - Monson	✓	
Rep - Carlisle	✓		Rep - Skarphol		✓
Rep - Delzer	✓		Rep - Svedjan	✓	
Rep - Glassheim	✓		Rep - Thoreson	✓	
Rep - Gulleson	✓		Rep - Warner	✓	
Rep - Huether	✓		Rep - Wentz	✓	
Rep - Kempenich	✓				
Rep - Kerzman	✓				
Rep - Kliniske	✓				

Total (Yes) 20 No 1

Absent 0

Floor Assignment Rep. Kliniske

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

2001 SENATE HUMAN SERVICES

CONFERENCE COMMITTEE

SB 2239

2001 SENATE STANDING COMMITTEE MINUTES

BILL/RESOLUTION NO. CC SB 2239

Senate Human Services Committee

☒ Conference Committee

Hearing Date April 13, 2001

Tape Number	Side A	Side B	Meter #
1		X	
April 17, 2001 1	X		
Committee Clerk Signature Carol Kolodejchuk			

Minutes:

The conference committee, SENATOR FISCHER, SENATOR KILZER, SENATOR MATHERN, REPRESENTATIVE PORTER, REPRESENTATIVE KLINISKY, REPRESENTATIVE CLEARY, was called to order by SENATOR FISCHER.

REPRESENTATIVE KLINISKY explained what the House amendments did to the bill. It moved the administration of the PKU program from the Health Department to the Department of Human Services. We did remove the funding from the budget of the Health Department. Human Services can absorb the cost so there is no appropriation. SENATOR MATHERN commented that he had no problem with the administration if it is properly funded. BRENDA WEISZ: We feel we can absorb the cost within the budget, but there has been no money moved into the department to cover this. The fiscal note is the total cost of running the program including those that are on the system right now and allowing for new cases that just came on and any potential cases. REPRESENTATIVE KILINISKY: We have a total of 22 on the program with one to be born any day now. On the high end we could possibly have 24 people on the program and this is built on 27 individuals. It was also built on adding food costs for children and they only receive formula. The cost of the formula was high and I'm very disappointed in the fact that it came out at $229,000 when this biennium we only spent $133,000. This is a move that we made; should not affect the families at all. If this passes, someone was told that children with cleft palates and heart disease would not be funded. This should not be used as pawns when it should not affect them at all. SENATOR MATHERN: What is the increase for? MS. WEISZ: One section talked about adults receiving food; reimbursing for food; clinics; responsibility without funds. SENATOR FISCHER: Where is the money that was available for this project. BOB BARNETT: PKU is included in Health Department budget in 2004. It totaled $213,977. That money was removed from the Health Department funds. The amount was $156,378 of special and Federal funds and $57,619 in general funds. This money was removed from Health Dept and I don't know where it is now. It should be included someplace.

Discussion continued. REPRESENTATIVE KLINISKY stated that this bill absolutely necessary and adding a fiscal note will kill it. Guidelines in the bill are vital. This program is mandatory. Discussion on fiscal note or appropriation on bill. The committee was recessed until 1012 is resolved.

April 17, 2001, Tape 1, Side A.

The conference committee was called back to order. SENATOR FISCHER reviewed the situation. Senator Solberg has amendments to include the funding in 1012 for $50,000 to the Human Services Department from the Health Department to administer the PKU program. SENATOR MATHERN asked if the House changed who would be covered.

REPRESENTATIVE KLINISKY: We did not change the bill except the administration to the

Department of Human Services. SENATOR MATHERN: Do you believe this amount of funding will cover the expenses of the program? REPRESENTATIVE KLINISKY: They said they needed $56,000 for the program and they would take $50,000. SENATOR MATHERN moved the Senate accede to House amendments. REPRESENTATIVE PORTER seconded the motion. Discussion ensued. Roll call vote carried 6-0-0. SENATOR FISCHER will carry the bill on the Senate floor; REPRESENTATIVE KLINISKY will carry the bill on the House floor.

Date: 4/11/01

Roll Call Vote #: 1

2001 SENATE STANDING COMMITTEE ROLL CALL VOTES
BILL/RESOLUTION NO. 2239

Senate Human Services Committee

☐ Subcommittee on

or

☒ Conference Committee

Legislative Council Amendment Number

Action Taken Senate Accede to House Amendment

Motion Made By Sen Mathern Seconded By Rep. Porter

Senators	Yes	No	Rep ~~Senators~~	Yes	No
Sen Fischer	✓		Rep Porter	✓	
Sen Kilzer	✓		Rep Kliniske	✓	
Sen Mathern	✓		Rep Cleary	✓	

Total (Yes) 6 No 0

Absent 0

Floor Assignment Sen Fischer / Rep Kliniske

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

REPORT OF CONFERENCE COMMITTEE (420)
April 17, 2001 2:30 p.m.

Module No: SR-68-8754

Insert LC: .

REPORT OF CONFERENCE COMMITTEE

SB 2239, as reengrossed: Your conference committee (Sens. Fischer, Kilzer, T. Mathern and Reps. Kliniske, Porter, Cleary) recommends that the **SENATE ACCEDE** to the House amendments on SJ page 533 and place SB 2239 on the Seventh order.

Reengrossed SB 2239 was placed on the Seventh order of business on the calendar.

2001 TESTIMONY

SB 2239

The Arc of North Dakota

P.O. Box 12420
2500 DeMers Avenue
Grand Forks, ND 58208-2420

January 25, 2001

Members of the Senate and House Human Services Committee:

The Arc is an organization committed to improving the quality of life for children and adults with mental retardation and related developmental disabilities and their families through advocacy, education and family support services. As an organization of 1,200 members, we would like to voice our strong support for the proposed Senate Bill #2239. This proposed legislation would ensure appropriate expensive prescription medical food to be provided **at no cost** by the State Health Department – Maternal Child Health (MCH) to males under the age of twenty-two and females under the age of forty-five diagnosed with Phenylketonuria (PKU) or Maple syrup Urine Disease (MSUD).

Additionally, to prevent large pharmacy mark-ups, this bill would allow the Health Department to offer medical food **at cost** to females age forty-five and older and to males twenty-two and older diagnosed with PKU or MSUD.

The proposed legislation would also ensure that the insurance companies continue to provide up to $3,000 worth of coverage for metabolic formula and low protein foods needed by PKU and MSUD patients. Mental Retardation and other severe neurological problems can develop if proper nutrition therapy is not maintained and life long treatment is necessary. The continuing coverage of prescription metabolic formula and low protein modified foods under the Department of Health – MDH programs and insurance policies diminishes the likelihood of developing mental retardation and controls the severity of mental retardation.

Equal access to care for all individuals with PKU or MSDU in North Dakota is highly desirable but personal financial barriers exist among the families. Specialized prescription medical food and low protein products are expensive and necessary to maintain one's well being and should be treated as medical necessities. Your support is needed on this bill, Senate Bill #2239.

Peter Johnson, Chairperson
Government Affairs Committee

January 26, 2001

To Whom It May Concern:

I am a pediatric doctor and specialize in endocrinology and metabolic diseases. I am the doctor that sees most of the metabolic diseases in the state, which include phenylketonuria (PKU) and maple syrup urine disease (MSUD). PKU is a silent disease. Patients who are not treated with a low protein diet and a special formula develop mental retardation, psychiatric problems, learning disabilities, and attention problems. Mothers who have PKU have babies that have small heads, mental retardation, and birth defects including heart defects. Children with PKU and infants born to mothers who have PKU need to be on a low protein diet. They also need to receive a special formula, which provides them with all of the essential nutrients that their body needs to grow and develop normally and also to maintain a normal pregnancy. Without this special formula, mental retardation and birth defects will occur. Maple syrup urine disease is a disease that if left untreated causes coma and death within a few days.

For many years North Dakota has been at the forefront of diagnosing and treating these conditions. If approved, the proposed Metabolic Food Bill will keep North Dakota at the forefront of child healthcare. Just one person that slips through the cracks will cost our state far more than the cost of formula for all 20 patients with these conditions. For this reason, I strongly support the approval of this bill, which will prevent these people from falling through the cracks.

If you need any further information about PKU, its treatment, or the consequences of not treating it, do not hesitate to contact me. There is only one right decision to be made in this situation. I hope that the Legislature continues to have enough foresight to make this important decision.

Sincerely yours,

Alan G. Kenien, M.D.

J
Lloyd W. Johnson
302 30th Ave. N Apt. 9
Fargo, ND 58102-1537

1-27-2001

Dear Sirs:

It seems that over the many years, I have lived in No Dak," 85 yrs except for 46 mos in WW II, North Dakota has been my home & Proud of No Dak, Our State has always taken Care of its own, even in So Called hard times; But it seems when it comes to our Citizens that have So called "Orphan Diseases" Some of our legislators want to quit Supplying said Formula to these citizens that need a little extra help, this would be a big hardship for a family to Pay.

For Cole Johnson family it Would be $700\00 a month for "Maple Syrup Disease" Control year in & Year out.

It seems that People who take Cure for addiction to Drugs & Alchol have a better chance of getting help, I read in Forum & Cash $150,000 to 200,000 to treat these citizens, $60,000 doesnt seem that much for 19 recipents to stay healthy, No one asked for this Problem.. its here lets take Care of it

Grandpa
Lloyd J.

January 29th , 2001

To Whom It May Concern:

My name is Dr. Todd Twogood and I am a pediatrician at Medcenter One in Bismarck. I also represent the North Dakota Chapter of the American Academy of Pediatrics (AAP). I stand before you today to voice the opinion and feelings of not only those who care for the children of our great state ,but, for the families of those who are here today and the families who could not be here. There is a great need for supporting the special diets and special formulas for individuals who have phenylketonuria (PKU), and maple syrup urine disease (MSUD). Without the financial support for the medical food necessary, there could be devastating effects. Mothers who have PKU could have babies born with small heads, mental retardation, and birth defects such as congenital heart disease. The special diet will help reduce this chance. Also babies born with PKU need the special formula and a low protein diet to grow and develop normally. Maple syrup urine disease, if left untreated causes coma and death within a few days.

North Dakota has been very supportive of providing for these individuals and families in the past. Newborn screening and financial support for the special diets will prove to be preventative, and ensure medical cost savings in the long run. I strongly support this bill and I speak for all the pediatricians of our state. I hope you also will support this important issue.

Sincerely yours,

Twogood MD

Todd Twogood, M.D., FAAP
Legislative Chairman and Sec/ Treasurer
of the NDAAP

January 29, 2001

SENATE HUMAN SERVICES COMMITTEE SB 2239

SENATOR LEE AND COMMITTEE MEMBERS:

My name is Jack McDonald. I am appearing today on behalf of The ARC of North Dakota.

The Arc 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 strong support for Senate Bill 2239. This proposed legislation would ensure appropriate <u>expensive prescription medical food</u> (formula) would be provided at no cost by the State Health Department – Maternal Child Health (MCH) to males under age twenty-two and females under age forty-five who are diagnosed with Phenylketonuria (PKU) or Maple Syrup Urine Disease (MSUD).

Additionally, it would allow the Health Department to offer for sale "<u>AT COST</u>" medical food to females age forty-five and over and to males twenty-two and order who are diagnosed with PKU/MSUD to prevent large pharmacy mark-ups.

The proposed legislation would also ensure that the insurance companies continue to provide up to $3,000 worth of coverage for metabolic formula and low protein use for PKU/MSUD patients. The continuing coverage of prescription metabolic formula (medical food) and low protein modified foods under the Department of Health – MCH program and insurance policies supports mental retardation efforts. Mental Retardation and other severe neurological and problems can develop if proper nutrition therapy is not maintained. Life long treatment is necessary.

Metabolic control in women with PKU who are pregnant or planning a pregnancy is crucial because of the serious effects of high phenylalanine (PHE) levels on the fetus. Microcephaly (small heads), mental retardation, and congenital heart disease are only just a few problems of babies born to PKU women who are not well controlled on the diet.

Equal access to care for all PKU individuals in North Dakota is highly desirable but current financial barriers exist. Specialized prescription medical food and low protein products are a medical necessity and should be treated as such. We urge your support on this important Bill.

If you have any questions, I'd be glad to try to answer them. THANK YOU FOR YOUR TIME AND CONSIDERATION.

North Dakota Dietetic Association

Senate Human Services Committee
1/29/01

The North Dakota Dietetic Association would like to go on record in favor of Senate Bill 2239, relating to in-born errors of metabolism. I would like to make a few key points for you today:

- Medical foods and low-protein modified food products are essential to the health and well being of those with in-born errors of metabolism.
- North Dakota residents with in-born errors of metabolism require frequent and specialized medical monitoring.
- Those who give birth at home must be made aware of the need to screen for early detection of and proper treatment of in-born errors of metabolism to reduce mortality, morbidity and associated disabilities.
- Medical foods and low-protein modified food products should be provided to those with in-born errors of metabolism as indicated in SB 2239. It is vital to children and women of childbearing age that they consume the foods provided for them to ensure proper growth and development for themselves and for the developing fetus. Making the medical food available at cost to adults is a way to help them follow a difficult, but necessary dietary regimen. Individuals who consume these food products as part of their overall dietary treatment will experience a better quality of life with fewer medical problems, and therefore, fewer medical costs.

The North Dakota Dietetic Association, a group of 291 nutrition professionals in North Dakota, strongly supports Senate Bill 2239. In-born errors of metabolism can have devastating results if treated improperly or not treated at all. North Dakota will provide an "ounce of prevention" with this bill, and avoid the expenses of the "pound of cure" associated with improperly managed in-born errors of metabolism.

Thank you for your time,

Karen K. Ehrens, LRD
North Dakota Dietetic Association

Testimony for SB 2239

Senate Human Services Committee
January 29, 2001

Madam Chair and committee members, for the record I represent Blue Cross Blue Shield of North Dakota. This bill will hopefully correct a problem resulting in a bill approved by the 1999 Legislature. That bill established a $3,000 insurance mandate cap. The intent of that bill was that the formula (medical food) was to be covered by the Health Department and those with insurance could use their insurance coverage for the low protein modified foods. However, by mistake, the way the bill was interpreted that there was only a $3,000 limit on low protein modified foods and no limit on the formula. This was not the understanding and agreement reached with the parents of children with a metabolic disease and BCBSND.

During the interim, the bill sponsors worked to develop a revised bill, which would provide free formula and still a $3,000 cap that could be used for the low protein modified food. The parents were very supportive and good to work with during this process. After several versions, the final version still provided a situation, which is unfair for those who have insurance. The essence of the bill states that the Health Department will provide formula free to men up to the age of 22 and women up to the age of 45. However, it states that those with insurance, the Health Department can collect the $3,000 policy limit to cover the cost of the formula. This version penalizes those who have insurance. Everyone else gets the formula free, however insured parents must use their insurance, thus adding to their lifetime insurance cap.

Sen. Fischer has proposed amendments, which will correct this inequity. His amendments will restore the original intent of the 1999 legislation. The Department will provide the formula free to all men up the age of 22 and all women up to the age of 45. In addition, parents can purchase formula from the department at cost in case there is a need beyond the established age deadlines. Those with insurance can use their $3,000 insurance limit for this additional formula need or for low protein modified foods. This gives the parents flexibility on how they want to use their insurance without penalizing them.

Madam Chair and committee members, BCBSND supports Senator Fischer's amendments and urge you to approve them and give SB 2239 a Do Pass As Amended. I would be willing to try to answer any questions the committee may have.

Dan Ulmer and Rod St. Aubyn
Government Relations
Blue Cross Blue Shield of North Dakota

Testimony on SB 2239
Regarding the Newborn Screening and Metabolic Food Programs
before the
Senate Committee on Human Services
by
Darleen Bartz, Department of Health
January 29, 2001

Good Morning Senator Lee and members of the Committee, I am Darleen Bartz, Chief of the Preventive Health Section of the state Department of Health. With me is Karen Oby, Newborn Screening Program Coordinator, for the Maternal and Child Health Division. I am pleased to present this testimony in support of SB 2239 which revises the newborn screening law and clarifies the Department of Health's medical food program and client eligibility for coverage of benefits.

In 1967, the legislature enacted chapter 25-17 that established the testing program for phenylketonuria (PKU) and other metabolic diseases. Under this law, the department was directed to make arrangements for necessary treatment if a family was unable to pay. For many years, the department has provided medical food to all families without regard to income.

In 1997, a new section was added to title 26.1 (North Dakota Insurance Code) which imposed a mandate on all health insurance companies to provide coverage for treatment. In 1999, this mandate was modified to make the Department the primary payer. The department was to commence a rulemaking process following this session to establish income eligibility for benefits. In May 1999 and again in March 2000 the department published proposed rule. Due to opposition, the Health Council accepted the Department's recommendation to suspend rulemaking and develop legislation that would clarify eligibility for benefits under the metabolic food program.

A workgroup, composed of 20 individuals was developed to examine this issue. This group included legislators with affected families in their districts, representation from the ND Medical Association including four physicians, consumer representation, an individual from the governor's office, health department staff, and an individual from legislative council. The consensus of the workgroup was that 1) PKU and Maple Syrup Urine Disease (MSUD) are significant public health issues and that medical food should be provided regardless of income; 2) insurance should be the first payer; 3) there should be a mechanism for the health department to collect from insurance companies, and others, and the funds go into the operating expenses for the department; 4) there should be a provision by which individuals who are not eligible for benefits can buy metabolic food through the department; and 5) the department would need additional funding to pay for this funding this treatment program. These areas are reflected in Senate Bill 2329 and the accompanying fiscal note.

In general, SB 2239 amends the newborn screening and metabolic food program to permit the Department of Health to make changes in the newborn screening program that incorporate advances in the technology of testing for metabolic diseases and the recommendations of the American Academy of Pediatrics.

The bill also gives explicit rulemaking authority to the Department of Health. Third, the amendments modify the law to reflect current practices and define more accurately the division of responsibility between Department of Health and Department Human Services for short-term and long-term follow up of children with special health care needs.

Due to the increased costs of medical food provided to individuals with PKU and MSUD and the increased program participation, it is projected that $80,000 will be needed during the coming biennium to continue to cover the treatment costs for all individuals with PKU and MSUD. If this bill is passed, making insurance the first payer up to $3000 per year for metabolic food and low protein food products, it is anticipated the department may collect up to $70,000. The remaining $10,000 will be needed from the general fund.

Attached is the testimony from Dr. Terry Dwelle, Chief Medical Officer for the Department. He is unable to be here today. In his testimony he provides information regarding these diseases and their significance as a public health concern.

In conclusion, I urge you to support this bill. Either Karen or myself would be happy to respond to any questions you may have.

Thank you.

Testimony on SB 2239
Regarding Newborn Screening and Metabolic Food Programs
before the
Senate Committee on Human Services
by
Terry Dwelle, MD, Department of Health
January 29, 2001

Good Morning Senator Lee and members of the Committee. I am Terry Dwelle, Chief Medical Officer of the North Dakota Department of Health. I am pleased to present this testimony in support of SB 2239 to revise the newborn screening law and the law establishing the Department of Health's metabolic food program.

My testimony addresses the reasons I am recommending, with Department of Health concurrence, support of providing medical food (formula) to all individuals diagnosed with PKU or maple syrup urine disease, without regard to income.

In critical metabolic diseases like PKU, there is an inverse relationship between age at treatment initiation, maintenance, and later IQ. The treatment of infants found to have PKU must commence no later than 7 to 10 days after birth and be maintained to at least early adulthood since brain development continues through adolescence. Fetal exposure to high maternal phenylalanine levels, particularly in the first trimester, is likely to produce microcephaly, significant and permanent mental deficiency and congenital heart disease. In these situations, it is not only critical to promptly identify the disease but also initiate and maintain this medical food intervention throughout the time of risk. Thousands of people who were screened as infants and treated successfully are now normal adults who likely can expect a normal lifespan, though more research is needed on the PKU and other metabolic disease processes in adults, with particular focus on the potential effects of aging.

Specialized medical foods and low protein products are a medical necessity for people with diseases like PKU and maple syrup urine disease. Low protein medical foods are expensive, costing 110 to 3,500 percent more than foods in a normal diet and are often only available via mail order from specialty suppliers. In my opinion, it is highly desirable to ensure children and families affected by these metabolic diseases in North Dakota provision of critical medical food during the high-risk situations of childhood and childbearing years in females, regardless of the social or economic status of the family. As we all know, income is not always an adequate indicator of financial solvency. It would be a great travesty for even one child in this great state to somehow slip between the cracks for whatever reason and develop irreversible mental retardation. The state support costs of just one child with PKU or MSUD developing irreversible mental retardation would far outstrip the minimal costs of underwriting medical food for these conditions.

Medical foods and low protein products are a medical necessity for people with metabolic diseases like PKU and MSUD and should be reimbursed by third-party payers, just as with other medically necessary drugs and devices for other diseases. The Health Department should be allowed to seek third-party reimbursement of medical food costs. To require assessment of third-party payment up-front could jeopardize the timely administration of medical food. In this light, it seems prudent to allow the Health Department to provide medical food without regard to social or economic status and then seek reimbursement.

For these reasons and considerations, I urge the Committee to approve SB 2239.

Madame Chairman, this completes my testimony. I would be pleased to answer any questions you or Committee members have regarding this legislation.

Good morning Madam Chairman, members of the Senate Human Services Committee my name is Brenda Rakoczy, I am the mother of Sydney who is 5 and has PKU, she is one of the many PKU success stories you see in this room. I want to thank you for the honor of testifying in front of the "People's Legislature" in **favor of SB 2239.**

[illegible] a rare metabolic disorder that causes profound mental retardation through the build-up of phenylalanine (PHE) in the brain and nervous system. If treatment begins shortly after birth, these affects can be avoided. As a result of state-mandated newborn screening and early and continued treatment, people with PKU can go on to lead normal lives and attain many of their goals and aspirations. PKU newborn screening has become compulsory since 1963 in the U.S. There has not been a child born with PKU in North Dakota in the last three years.

Without SB2239, several barriers (mostly financial) will remain in preventing and treating this disease among our state's children and young adults. Quite simply, the treatment of PKU is one of the most economically valid and morally responsible investments we can make in our society. Thus the need for special metabolic formulas (medical food), low protein modified food and a way to pay for it – will remain throughout a PKU individual's life.

If individuals 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. Following a very restricted low protein diet can prevent this terrible development; this regimented diet restricts all meats, dairy, poultry, legumes, nuts, and most products containing flour and grains. A typical diet includes preciously weighed fruits and vegetables low in protein (phenylalanine).

Doctor Levy, Children's Hospital – Boston, stated: "Generally as a result of such a strict diet so low in phenlylanine, so low in protein that a person without PKU would go blind or either die. But it doesn't necessarily [illegible] what most of us consider a healthy diet of fruits and vegetables and salads. These also contain phenylalnine. [illegible] with PKU vary in their ability to process phenylalanine. (PHE) Some can get away with eating salads or [illegible]ven pizza with no cheese or meat. For others, however, four lettuce leaves, or an ear of corn is an entire day's quota of PHE." (As very similar to my daughter's Sydney's allowance – only 275 mg per day, as she grows her tolerance does not necessarily increase it can stay the same even when she is 40).

The heart of the problem now encountered by a family whose child is diagnosed with PKU is no insurance or lack of appropriate coverage (due to self funded plans and federal insurance plans) and possibly severe financial cuts to the formula program from the Health Dept if this bill is not approved. On behalf of the PKU families we would like it noted on legislative record that the true legislative intent of SB 2374 which was heard last legislative session was to have a combined cap for both medical food (formula) and low protein modified foods at $3,000 total. While it would have been great to have no fiscal limit, we feel it is important to be honorable and have the capped instituted as was originally testified to during the last two legislative sessions.

As a PKU individual or child grows so will their intake of formula. Sydney's prescription metabolic formula cost is currently $152.00 for 2 cans, she goes through approximately 8 cans per month at $608.00, than the metabolic flavoring to disguise the terrible taste of the formula is $46.00 for 6 cans, she uses approximate 5 cans per month. Low protein modified foods run approximately $50 - 100 per month. Keep in mind she is only five years old. Sydney prescription medical food not including her low protein modified food is over $7,296 per year. I'm honorably retired from the Air Force and Tricare is a federal insurance company and does not cover the prescription medical formula or low protein modified foods. Sydney's monthly medicine expense is higher than our home mortgage.

[illegible]out assistance from the state health department, we would have a very difficult financial barrier to providing [illegible] her immediate critical medical needs. Without passage of this bill one of these ND children will fall through the cracks. **Off-diet PKU/MSUD individuals are literally lives in danger.**

The cost of metabolic formulas are minuscule compared to the cost of housing and educating a child with mental retardation which is approximately $100 -- 125,000 per year.

I would like to personally thank the Health Dept for providing the medical formula for the families over the last 30 [illegible] North Dakota is a forerunner in treating and preventing mental retardations in our state's most precious [illegible]ces – our children. I would also like to sincerely thank the Health Dept, Sponsors of this bill and medical officials who took part in working on this vital piece of legislation. Additionally, I would like the thank the insurance companies for covering up to $3,000 dollar for low protein modified foods for individuals with true insurance coverage and for those individuals who are over the age guidelines for benefits from the Health Dept. Combining our efforts – we can ensure no individual falls through the cracks. It is too high a price to pay.

During the 56th Legislative Assembly - House Human Services Hearings the State Health Dept testified and we PKU families agree with their position that the purpose of the Department's Program - is to avoid the cost and burden of severe mental retardation. "

Additionally they testified, "The Department will be able to maintain a register and assure that all eligible individuals, particularly children, receive necessary formula and low protein modified food products. If insurance companies had this responsibility, those with metabolic conditions could be subject to high deductibles or co-payments, and the Dept would not have the 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.

The Health Officer went on to testify that individual would move on and off health insurance coverage or change policies under which they have different co-payments and deductibles that could disrupt the continuity of these special dietary supplements." Also 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 risk of mental retardation."

[illegible]onally, as stated in his testimony that the Health Dept is in the best position to ensure individuals get their formula without any interruptions. The state of ND 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 Dept of Health has provided formula and low protein foods that have helped many families and children over the years."

It should be of special note – the Health Dept has provided medical formula for families who do not qualify for Medicaid. Individuals who get their formula from Medicaid – not paid from the Health Dept MCH Block Grant receive some low-protein modified foods. The rest of the families who get formula through the Health Dept do not get low protein modified foods they are appropriated through personal income our through an individuals insurance company if they have appropriate coverage.

We PKU families certainly support this position. One additional note that is vital in this bill is individuals with PKU/MSUD over the age limits be authorized to purchase the formula directly through the Health Dept "AT COST." This will save astronomical mark-ups from pharmacies as the $3,000 cap on insurance will be vastly eaten up and those without insurance will have to pay the full cost out of their pockets. This is a tremendous attribute of this bill.

In closing, I would like to tell you how humble I am to have to come ask for your assistance. As parents we always envision when our children have a need, we will always be able to take care of it. Sadly, this is one thing that I cannot be my child's "all and all" – I need your help. **Please vote yes on SB 2239.**

[illegible]renda S. Rakoczy

[illegible]ery Respectfully – Brenda and Tom Rakoczy

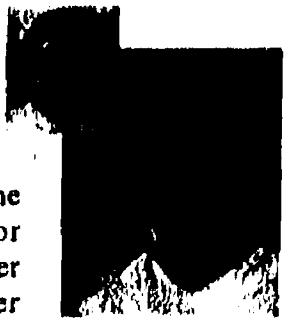
Special Features

[illegible] follow-up to the research report featured on page 3, the three stories here show first-hand how even late-diagnosed adults with PKU can benefit from the PKU diet—at any age. In the process, they remind us all of how fortunate are our children who are diagnosed and treated from infancy.

Mickey's Story

by Emily Farmer, Dexter, ME

My sole objective in writing this story is to give courage to everyone preparing and consuming PKU diets. My severely retarded daughter, Michelle (Mickey) Farmer, was born in a small hospital in Maine on Nov. 26, 1956, a time when there was no routine PKU test done for newborns. A year later our second daughter (non-PKU) was born.



Mickey Farmer, 1958 and today.

As new parents, it took us awhile to recognize that Mickey's sister was surpassing Mickey in all forms of mental progress. When Mickey was twenty-two months old, we took her to a pediatrician in a city near us. He felt she might be deaf or have cerebral palsy. He made an appointment for us at Children's Hospital in Boston. It was there that we learned Mickey had PKU. However, in talking with the clinic specialist, we were told that it was too late to place Mickey on a PKU diet. His exact words, which I have in a letter dated December 4, 1961, are these:

> *"I still would stand on the opinion that a phenylalanine deficient diet would not improve Michelle's intelligence to the point of being teachable. It might, however, improve her behavior and if you find this sufficiently difficult at home to warrant the unavoidable difficulties in maintaining such a child on a very unnatural diet, it would not be medically wrong to give it a try. I should not personally advise it, but other physicians might well feel differently and it would be reasonable to try if you can find one locally who is prepared and qualified to carry out the close medical supervision necessary."*

This idea that the diet probably would not help our daughter was reinforced a number of times—until we believed it. But since subscribing to National PKU News and the PKU Listserv group, I now have a great deal of regret that we did not place Mickey on the diet despite the advice of the physicians. I truly believe that she (and we) would have done considerably better.

Early on, my husband and I and two other couples formed an organization made up of parents of mentally retarded children. Our organization built a school for our small community, funded by federal and county money. One fortunate thing is that Mickey has always been able to attend a day program. The following years were trying though, with many difficult behaviors to contend with. All of this was tolerable until Mickey was about twenty years old. Then, her behavior really got out of hand. We made an appointment with Dr. George Jervis (a specialist in PKU, now deceased) at Letchworth Village in Thiells, New York. He examined Mickey and talked to us about trying to cut down on her phe intake, but not necessarily going on the full diet. I tried the diet, but with little help and not much faith in myself. We also had a doctor living next door who gave me prescriptions for a number of drugs to see if they would help. I hated the effects these drugs had on Mickey. I did not feel any of it was working in the slightest. I was very discouraged.

When Mickey was twenty-three, we placed her in a new facility that opened close to our home, because of her behavior but mostly because there was to be a registered dietitian. They started the PKU diet, but gave her too much phenylalanine and her blood levels stayed between 12 to 20 mg/dl, too high for many positive changes. She did not do well there. It was very close quarters and a restrictive environment.

We were delighted when Mickey had an opportunity to move to a group home. There the cook prepared the menus and had them approved by a dietitian. The meals were an improvement but still not great. Her blood levels ranged from 10 to 12 mg/dl. Her behavior improved somewhat but still she was difficult. (Mickey is nonverbal for the most part and her frustration level is high. She also has autistic tendencies due to PKU.)

In December 1995, Mickey moved to a home with just one other resident and staffed around the clock. We were thrilled. There was no dietitian assigned, though, and the staff was not knowledgeable about the PKU diet. When I got tired of the repetitive menus being offered and Mick's high blood levels, I spent summer 1996 working up a five-week menu. Now, since my diets (approved by a dietitian) have been used, Mickey's blood levels have ranged from 3 to 8 mg/dl.

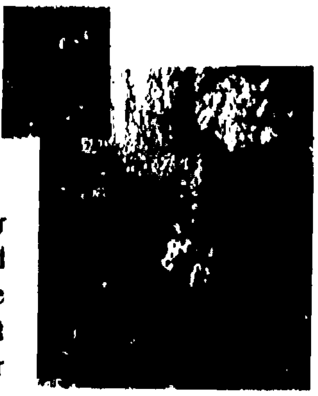
Between the lower blood levels and continuing 2 mg. of Risperdal a day, Mickey has shown very significant improvements. She is a much happier person. She has never been at all sociable, but that aspect has improved a great deal. She actually enjoys a limited number of outings and can indicate, either by signing or verbally saying yes or no, when she does or does not wish to participate. She is much more pleasant to have around. There are still ups and downs, but we are so very pleased with the progress she has made. It has been a lesson in perseverance. I will not give up on trying to make Mickey's life as comfortable and rewarding as possible until the day I die.

When you get discouraged, just think of Mickey. It should give those of you with early-treated children the courage to continue the every day struggle and realize just how necessary the effort is!

Cathy's Story

by Francis Polansky, Irving, TX

My daughter with PKU, Catherine Ann, age 53, is profoundly retarded. She has a measured I.Q. of less than 20. She has lived at the Denton State School in Texas since she was 16. Cathy was 11 years old before her PKU was diagnosed, after all those years of our frustration and heartache. The low protein diet we started for her at that time was stopped after a year, on the advice of her pediatrician. He said that the damage to her brain was irreparable. We were devastated. It was 1956.



Cathy Polansky, 1951 and today (mom on right).

Cathy's development was very delayed. For example, she did not walk until she was 27 months old. Eczema and erratic aggressive behavior made her life miserable. She would pace in one corner of a room, never interacting with her peers. She would butt her head against a wall and pull her hair out daily. After we placed her in the Denton School, managing her constant agitation was a great challenge for her caretakers.

In 1982, the Denton School started a one-year diet trial with 7 PKU residents. Cathy's eczema improved, but the project proved to be impossible due to limitations in a state institution where 32 residents had only 2 caretakers. So large doses of psychotropic

Senate Bill-Metabolic

TO WHOM IT MAY CONCERN:

RE: Senate Bill Number 2239
Metabolic Ruling

I will try to keep this short. Our son, Cole, was born with a rare genetic disease called Maple Syrup Urine Disease (MSUD). It affects about one in 300,000 births. Cole is the only child diagnosed with this disease in North Dakota is the past nine years. This disease affects the way the body processes certain components of protein. 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 interferes 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 usually dies by 14 days of age. Cole was a very lucky baby, he was in a coma for five days before being diagnosed at 16 days of age. Doctors didn't know if he would have irreversible brain damage. But we have been blessed. Cole functions like any normal nine year old.

Almost any illness Cole suffers can become life-threatening. He has been hospitalized eight times. The most serous was at age 3 1/2. Cole had the flu and couldn't drink his formula. Dr. Alan Kenien put him in the hospital for his normal treatment. After a couple of hours he started seizures and went into a metabolic crisis with severe brain edema. Thanks to the quick work of Dr. Kenien and Dr. Holmes Morton from Lancaster, Pennsylvania, Cole survived. Cole's last hospital stay was three weeks ago with walking pneumonia.

Cole needs his special formula and low protein foods to survive along with the right medical care. Without these we would not have our son.

We are very grateful to the State of North Dakota MCH Division. And we pray that it will continue. Without the state's help Coles formula would cost $8,400.00 a year, plus the cost of low protein foods. This formula would cost us at least $700.00 a month. How would you like to live with the feeling, if you can't make your son's formula payment he will die?

Thank you
Sincerly

Jeff and Amy Johnson

My name is Anne Christians. I am a 24 year old phenylketonuric. As a child I needed my blood drawn frequently to monitor my metabolic disorder. I always wanted to be the one to draw the blood. Today I do. I graduated from NDSU with a degree in Clinical Laboratory Science. I am currently a medical technologist at a Fargo hospital. Without my formula this would have been impossible. Not only has the PKU diet guaranteed me a "normal" life but without it I would have severe mental retardation. I have been on this 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. The formula I drink provides my body with all the nutrients that I can't get through my diet. 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 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. It is a fact that PKU woman, not on the diet, give birth to children with mental retardation even though these babies are not born with the disease. This is because the baby gets its nourishment from the mother and if the levels of Phenylalanine are elevated in the mother, the baby's brain will not develop during pregnancy. As this is a genetic disorder, I will probably not pass on PKU to my children. I would however, have to be on this diet throughout my childbearing years to be able to have normal healthy children.

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 pass Senate Bill No. 2239

Madam Chairman Senator Le, and members of the Human Services Committee.

My name is David Aakre, I live in Kindred, ND with my four children, Danielle, DeAnna, Dolan, and Dalton. Danielle, Dolan, and Dalton are diagnosed with, and are being successfully treated for the metabolic disease called Phenylketonuria or PKU. DeAnna does not have PKU. Looking at them you can not tell who is diagnosed with PKU, and who isn't. If however the children with PKU were not treated you would have no problem telling who they were. They would likely be in wheelchairs, unable to control their actions, and incapable of engaging in normal conversation. From birth this disease silently, and slowly renders the individual with PKU to a totally dependent state of care and support if left untreated.

This is my third time here at the State Capital to take part in the legislative process, and it's still an awesome place. I can only imagine what it's like to be here on a daily basis, and what the feeling was when each of you first took office. There are some similarities between what you experience here, and what we as parents of the children with metabolic diseases experience. The same feelings you have when you introduce new legislation, and have success in seeing it through the legislative process is just like us. These are milestones in your career, and is how your effectiveness is measured. You see when our children reach milestones in their life such as smiling, rolling over, sitting up, walking, talking, starting school, participating in school activities, graduating, and leading productive lives, is where we measure our effectiveness in the treatment of these diseases. At certain points in their lives it is expected that they can perform, or be able to do certain things. When we reach these milestones there is much rejoicing, and a wonderful feeling of accomplishment.

This bill guarantees the continued success for the treatment of these metabolic diseases, and will ensure these children, and adults will have a chance to succeed in life. The consequences of having just one child not treated for this disease would be heartbreaking, and costly. This was the experience when the Guthrie test was first instituted. Children born just a few months prior to this were not treated, and consequently suffered irreversible neurological damage. The cost induced for the care of one individual not treated for this disease is much greater than the cost required to treat all the children, and adults in North Dakota. This is the testimony for the successful treatment of these metabolic diseases, which this [illegible]

Danielle recently has competed at speech events in the inform category, and chose PKU [illegible]. She placed fifth her first time participating at a speech event at the Valley City Optimist speech tournament. Parents are always proud of their children, as I am with mine, but for the parent of a child with these metabolic diseases it adds a generous amount of thankfulness to see them succeed.

Thank you very much for your prayerful consideration for this legislation. I think Danielle is going to go right into her speech so I'll introduce her. Here is my daughter Danielle Aakre.

David

David C. Aakre
P.O. Box 84
Kindred, ND 58051

PKU Speech
writted and given by
Danielle Aakre, PKU Patient

Imagine that you have just become the mother or father of a newborn baby. When your baby was born, a prick in the heal would determine whether or not your baby has a rare disorder. This test is called the Guthrie test.

This may not matter to you right now but if one day you want to become a parent, your baby will have this test done and the out come may mean a signifacant change for your family.

I f this disorder Phenylketonuria also known as PKU is not diagnosed at birth or treated correctly, the results will be very devistating.

Today I am discussing what PKU is, the consequences of being misdiagnosed or not being treated properly, and how it can be managed so the PKU patient can live a happy, normal life.

PKU is an inherited condition that can cause mental retardation if it is not treated properly.

PKU was discoverd in 1934 by a Norweigian, Dr. Asbjorn Folling. A mother of two retarded children went from doctor to doctor explaining to them the musty odor that her children were expiereincing. Dr. Folling became intrested in her children He found that these children had an abnormal substance in their urine. He studied other children and those with the same substance in their urine were diagnosed with PKU.

PKU is caused by a defect in a gene. Genes are present in every human cell. They give the cell information about its job. In PKU information is missing that tells about making an enzyme to metabolize protein. This enzyme is made in the liver and its called phenylalanine hydroxylase. Chidren with PKU have too little or none of this enzyme.

This disorder is very rare, about one out of every 10,000 babies have this disorder. It effects people of all ethinic groups but is more common in people of northern European descent.

If PKU is un diagnosed or is not treated properly the PKU patient could live a very devistating life not only would the child suffer but so would the rest of the family. The PKU patient can suffer mental retardation, which will prevent them from doing anything on their own, microcephaly which is an abnormal sized head, seizures, eczema which is a skin rash, and hyperactivity so severe where one cannot control their actions. This all happens because of the build up of protein in the blood and body tissue, and then goes to the brain and kills brain cells.

PKU patients are immediatly put on a low protien diet at birth. The amount of phenylalanine is determined for each child based on the phenylalanine blood level. The diet includes a special formula. This formula provides vitamins and minerals. It is recomended that PKU patients stay on their diet for life.

There are two kinds of basic formulas that provide the child with needed protein. They will either have little or no phenylalanine. Formulas for older children, teens, and adults contain no phenylalanine.This is so that they can have the flexability to have other foods. The formula for babies contains phenylalanine, because babies cant eat foods. Unfortunatly, because of the smell and taste of the formula, parents have to be very careful not to make faces, because that will discourage the children from dirinking it.

Foods people with PKU must eliminate are meats,fish, dairy products, nuts, and legumes. The diet includes formulas, sweets, veggies, fruits, fats, and limited amounts of grain products.

The severity for PKU varies in large amounts.There have been 192 diagnosed variations so far.Normal phenylalanine blood levels are less than 2mg at birth, at diagnosis most chidlren with PKU have high phenylalanine blood levels, typically they will be 29 mg or more. The amount of phenylalanine PKU patients are allowed vary as well. Some PKU patients are allowed 500mg of phenylalanine, while others can only have 50mg or less. For an example: Today you walked into Mc Donalds and bought a hamburger. For most PKU patients that will be all they are allowed to eat for the entire day.

Today I have discussed with you the consequences of PKU being misdiagnosed or not being treated properly how PKU can be managed properly, so the PKU patient can live a happy normal life.

There is a path to take for PKU patients that will not include the suffering, and will give the opportunity to live a happy normal life.

Hopefully you as the parent of a beautiful newborn baby have no concerns and do not have to deal with the disorder, but even if you a have a PKU child you know that your child can lead a happy normal life if it is diagnosed and treated properly.

Danielle Aakre

Bill #2239
Nancy Barrett Ouderkirk and Evan Ouderkirk
1-29-01

My name is Nancy Ouderkirk and this is my 5 year-old son, Evan. It has been five years since we first stood before you on behalf of North Dakota's PKU and MSUD children. And even though you may not remember us individually, you've had the opportunity to witness a success story.
When we were here initially, Evan was an infant. A great deal of uncertainty laid ahead for him. However, during these last 5 years, not only I have learned to better handle his disorder but so has Evan. Evan helps to make his own formula most mornings. He counts 17 tablespoons as I pour them into the blender and he mixes it. He helps mix up his own pancakes. Evan also rolls out his own tortillas and chips. Evan gets his own snack for school and puts it in his backpack. He even teaches me to play some of his computer games. Aside from Buzz Lightyear being one of Evan's favorite TV shows, he also likes to watch the History Channel. On numerous occasions Evan has enjoyed telling anyone who will listen about the Battle of Williamsburg and Haunted History. Evan is in preschool. He also is in the preschool choir at our church. He picks on his sister and plays with his friends.

Evan may not be seen as a remarkable child. He may seem like any other normal 5 year-old boy. Thank God! And thank God that the majority of other children don't have to go through what these kids have to go through to remain normal. These young ladies and gentlemen are all success stories. And for this reason I wish with deepest sincerity to thank all the professionals who have worked with our children, all of you, the Health Department, the Insurance companies, and the parents. It's because each one of these entities has worked in the best interest of PKU and MSUD children that we're all success stories. Because of all of us none of these children have fallen through the cracks or worse, had to be institutionalized. It is our hope that the current assistance we receive from the state will continue. With that having been said, please don't think that we are expecting the entire concern for our children's health be shouldered by the Health Department. We as parents very definitely do our part.

Aside from the formula, the majority of Evan's dietary needs are met by what I can purchase from the grocery store. However, being Evan can't eat any meat products, dairy products or any high protein food items, this also means he can't eat anything containing flour, eggs, soy or nutrasweet. As you may have noticed this greatly limits what he can eat. Therefore, it is necessary that I special order additional foods for him.

I recently ordered 6 food items for Evan – 4 boxes of pasta, one box of rice, and one box of chocolate flavored candy. The bill came to $50.30. That averages @ $8 per box. Out of necessity Evan is a very strict vegetarian. He eats low protein pasta; low pro breads and we make our own pizza crust. In addition to daily monitoring of Evan's food intake, there is the preparation of the food. I'm sure that when you are at home preparing your meals, most of you only prepare one meal per sitting. However in our house, at most meal times I must prepare two main courses. If we are having macaroni and cheese, I prepare Evan's special ordered cheese and his pasta. And then I make regular macaroni and cheese for my daughter and myself. Doing this for one day is no big deal. But continuing this day after day requires a great deal of organization and forethought. Even though Evan can eat fries from Mc Donald's, there's @ 128 mgs. of phe per small serving. That's over half of his daily diet and doesn't usually fill him up. And he can no longer eat their salads. Consequently, we don't go there often. The closest thing to fast food Evan really has is what I make ahead and freeze. Going on a family vacation means that we take it all with us because we most likely can't buy it when we get there.

Even something as simple and fun as Halloween takes more thinking ahead than the average family. We want Evan to be as much like any other child as possible. We don't want PKU to ever be a reason for him to not participate. That's one reason why every year at Halloween I prepackage candy items and I deliver them to each of the houses in our cul-de-sac so that when Evan rings their door bell he will receive something just like every other child who goes to that door.

Evan's current dietary needs cost approximately $4700 per year. Each case of formula costs the Health Department about $160. Evan uses @ 2 cases per month which is $320 per month - $3840 per year. His special ordered low protein food costs are approximately $70 per month - $840 per year.

We strongly hope you will support Bill #2239 as it represents a <u>guaranteed</u> program. It's a <u>guarantee</u> that if these children are provided with their formula (and ingest it) that they will remain mentally healthy. The other side of the coin is...that if these same children do not receive their formula, it is <u>guaranteed</u> that they <u>will</u> experience mental retardation and the cost emotionally and financially for the parents, children and the Health Department will be much greater. By voting in support of Bill #2239, you will guarantee continued success for the PKU and MSUD children of the state of North Dakota. On behalf of Evan, his sister Kailynn, their father and myself we thank you for your consideration.

We are the very proud parents of three beautiful daughters. All were considered very healthy and happy. Torie is 18, Tera 14, and Candace 8. My husband works as a Supervisor for the Falkirk Mine, and I work part-time for the local bank. I don't like writing things like this, because I'm not that good at it, but this concerns my number one thing in my life, and that is the health of one of my children.

Tera, our middle child, was born with a metabolic disease called PKU. I remember the day I was told this like it was yesterday. We were both devastated. We, like you, asked what was this disease, and will our little girl be all right. Well, we had a lot to learn. The only thing I heard as a mother was if this baby were not on this special formula, followed by a very strict diet, she would become mentally retarded. I could not believe that just diet would prevent this from happening, but we were both ready to do all we could.

We had a lot to learn. The formula, that this baby would have to have, could not be purchased from your local grocery or drug store. You could only get it from the State Health Department. We were told then that this formula was very, very expensive. We were so relieved that it was covered by the State, because there is no way we could afford the cost of just milk for this baby. As Tera grew older and food was coming into the picture, it was again a struggle. Everything that went into this child's mouth had to be weighed and measured. We took one day at a time. We had to. Carefully measuring, and making sure she drank all of her recommended amount of formula, and ate just the right amount of foods. When I say foods, it wasn't much food. The formula that these babies and children drink is such an important part of this diet. That comes first. You don't know how many times I've said, **"Drink your formula"**. It is so important.

Tera is now in the 8th grade and is a very active teenager. She is in basketball, cheerleading, dance, plays the flute in the school band, and runs track. She does well in school and is very well behaved and is liked by her peers. She takes her lunch to school every day. Pizza parties, birthday parties, and outings with her friends are a lot of planning. She can't have a slice of pizza or a hamburger, or for that matter, a lot of foods. We have to make hers special. Tera handles her diet very well, for the most part. Peer pressure is hard enough to deal with, let alone have something just a little bit different than everybody else.

Tera understands her diet, and the high price she would have to pay, by not being on it. She drinks her "so important" formula daily. It tastes and smells terrible, but it has all the vitamins and minerals in it that she doesn't get from the foods that she can't have. We often tell her she is like a diabetic and the formula is like insulin. It's her medicine. I look at Tera today, and just can't believe that we have come this far. This diet does work. Just look around the room at these healthy children. It's because of the formula and diet. They could have all been State cases. What I mean by that is, "institutionalized". And here they are!! When I say proud, we are very proud of what Tera has accomplished so far in her life.

Tera would like to, someday, get married and have a family, that is, after college. She would have to still be on this diet, as we are told "for life". If she were not on this diet of formula and carefully measured out food, babies would not be a good thing to have.

We have worked so hard for 14 years of her life to do everything right. Please don't take her health away from her. The cost would devastate us as a family.

Like I said, I am not a good writer, but I do take pride in being a good mother. By any means, we are not looking for a handout. We pay our bills, and support our city and state. We are just asking you to really look at this with an open mind.

Thank you.
Kim & Brenda Ankenbauer
Parents of Tera Ankenbauer

Formula $372./per month
$4464./per year

My name is Renee Christians. I'm a native North Dakotan and live in Fargo. I am the mother of two young adults with PKU. My children, Anne, age 24, and Daniel, age 22, are the oldest PKU children in the state who have been on this restricted diet since birth. Anne has graduated from NDSU and is currently employed as a medical technologist at a Fargo hospital. Daniel is a senior at UND and will be graduating in May.

This is the seventh time that I have testified about PKU. Today I would like to speak to you from the heart. Twenty-four years ago, I got a from call from my doctor telling me that my daughter had PKU and that she would have to be on a restricted diet and drink a special formula. When I asked my doctor what would happen if we didn't do this, he said that Anne would be severely mentally retarded. I was stunned and shocked. I wanted guarantees that if we followed this diet Anne would be fine. All I got were maybes, probablies and chances are good that she would be fine. I remember picking up my baby and holding her most of the day and crying. Until the new formula came, I felt that every bottle of I fed her was poison. When the formula arrived, I quick mixed it up and couldn't wait to feed Anne. She wouldn't drink it. She would suck a few times and spit the nipple out. By the end of the day I was frantic and Anne hadn't stopped crying because she was so hungry. When I tried to feed her yet again, I first tried the formula on my wrist but nothing came out. This formula was so thick it was plugging the nipple. She hadn't been able to get anything but I didn't know it. I fixed the problem and everything was fine. That was day one. For years I measured all the food I put on their plates. When they were done I would measure what was left. I sent weekly reports to their dietician. Over the years they have been poked and prodded with blood tests, physical exams and mental exams. We have dealt with weaning them from a bottle, school lunches, birthday parties, and the teen years of wanting to be just like your peers. Through the years, some of the struggles and challenges have been great but the rewards have been greater. The point I am trying to make is that while we had many worries, the one thing we never had to worry about was the support of the Department of Maternal and Child Health and the wonderful people who work there. They were there for my family. I never gave the formula a second thought, when I needed more they sent it. When I had questions they answered them. When I was scared they reassured me. I thank them for letting me spend my time and energies raising two wonderful and healthy children. On behalf of all the other PKU families, I ask you to let us parent and to not have to spend our time worrying about the financial aspects of this. Sometimes the expenses are so great that some families would not be able to afford this diet. The states return on their investment is the health of PKU children is profound. This is a slam dunk. On the diet, these children grow to be normal intelligent adults.

At this time, my son is no longer covered under the Department of Maternal and Child Health. What we would like to see happen, is that young men be able to continue to order their formula from the health department at cost. The state health department would then bill their insurance companies up to $3000, and in our case, my son for the difference. We have been ordering formula through a local pharmacy in Fargo and the markup is substantial. The Maximum XP is $50 per can at cost and $85 per can with the markup. I told them this was ridiculous but they said they had bills to pay too.

My daughter would fall under the Department of Maternal and Child Health with respect to future pregnancies. We would like to see the state continue to supply formula for woman of childbearing years up to the age of 45. In the case of my daughter, she is now employed and has her own medical coverage. The state would then bill her provider for $3000 with the state health department paying the difference.

When the answer is so clear, why would be take the chance of not providing these children with means of obtaining this formula. Please, I urge you on behalf of myself, my children and all the other PKU families to pass Senate Bill No. 2239.

January 29 2001

Madam Chairman and members of the Senate Human Services committee my name is Angela Maley I am the parent of Kayla Buchholz. Who is one of the few people in North Dakota that has been diagnosed with PKU. Kayla is a very beautiful healthy 7 year old. At this time Kayla is doing very well, thanks to her being able to receive her prescription metabolic formula and her low protein metabolic foods.

As a single parent and relying on my income I feel I would not be able to afford the necessities that Kayla needs to have for a healthy normal life.

My concerns as a parent with a child that has PKU, if the funding is to be cut I am afraid that Kayla would be one of the unfortunate ones that wouldn't get the necessities she needs and that she wouldn't be able to have the opportunities in life like the rest of us. I would hate for her not to get that opportunities in life because I wasn't able to afford. I wouldn't be able to forgive myself.

Please give my daughter Kayla Buchholz a chance at having a normal life and take all this into consideration and vote yes to SB2239.

Sincerely,

Angela Maley

To Whom It May Concern:

I am the mother of a beautiful, healthy 4 1/2 year old daughter named Kaylin Jo who has a metabolic disorder which is known as PKU.

Kaylin was officially diagnosed at 7 days old after a simple blood screening was done at birth. This simple test, which the state of North Dakota requires, may have saved her from a life of severe disability and saved taxpayers thousands of dollars. Kaylin was immediately prescribed a metabolic formula from a local pediatrician in our area, Dr. Lunn. She was also referred to a specialist in Fargo, ND whom had extensive knowledge in metabolic disorders, Dr. Kenien.

Kaylin began as an infant drinking the metabolic formula and has done very well with it. She, even to this day, loves her "milk". My husband and I have been fortunate in that we have not had problems getting her to drink her formula and therefore she has grown "like a weed". She currently is in the 95% for growth which in my eyes must mean we are doing something right.

As you may or may not know the formula is a staple in PKU children & adults diets. We have been told that this formula is extremely costly if we were to purchase it ourselves and are so thankful that the state pays for and is able to obtain the formula for us.

My husband and I both work out of the home full-time and still struggle as many families to do to make ends meet. We have 3 small children therefore have many additional household expenses including daycare. If we were required to purchase the formula ourselves I fear many of my children's other needs may go unmet.

Kaylin is fortunate in the fact that her body seems to be able to process a substantial amount of phe per day. We have been told that her formula may contribute to this. She typically requires about 600 mg per day but has been given up to 800 mg per day with good blood test results. My husband and I are also fortunate because this means we don't have to purchase as much low protein foods as other PKU families do. Kaylin is able to eat "normal" food which is low in protein and occasionally some specialty low protein food.

Senate Bill 2239 is important to our family because even with our health insurance coverage we would have a large out-of-pocket expense for formula in excess of what our health insurance would cover plus approximately $500 a year in low protein foods that must be purchased and shipped to our home.

Please vote "yes" on this Bill.

Thank you,

Tammy J. Anderson
Mother to Kaylin Anderson
Manvel, ND

TESTIMONY: NORTH DAKOTA CHAPTER OF THE AMERICAN ACADEMY OF PEDIATRICS

Eric R. Lunn, MD
Fellow American Academy of Pediatrics
Senate Bill #2239
January 30, 2001

Madam Chairman, Members of the Human Services Committee:

Thank you for the opportunity to provide written testimony today. I sincerely apologize for being unable to testify in person; however, due to my clinical practice, I am unable to travel to Bismarck at this time. I am truly sorry for this inconvenience. My name is Eric R. 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 is an international organization who not only provide health care for children but share a long-standing deep commitment to be the voices for innocent children throughout the world. I also appear in my own right as a concerned pediatrician to express support for Senate bill #2239.

Since the 1960s, the North Dakota Department of Health has conducted a metabolic screening program for newborns. This screening program identifies any child who may have a rare metabolic disease. It is critical to identify children with these disorders early in life so that treatment can be instituted shortly after birth. Children with these metabolic diseases that are left untreated generally progress to severe mental retardation and death. However, if treatment is begun shortly after birth, mental retardation and many of the other consequences of untreated disease can be prevented. Treatment for the various disorders might include restriction of one or more amino acids in the diet, restriction of total protein in the diet, or supplementation of specific substances such as vitamins. Again, untreated disease results in severe mental retardation or death.

Identifying a child with a rare metabolic disease is only half of the battle. If the child does not receive appropriate treatment, mental retardation will ensue. Therefore, it is critical for these individuals to receive proper treatment throughout their life. 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, or eggs. Special medical foods, including low protein and modified food products, are indispensable for the active, ongoing treatment of these diagnosed metabolic diseases. Not only are these diseases an emotional hardship for the family, it is also a financial hardship because of the costs of specialized foods. The estimated cost of supplying special food to a child with phenylketonuria (PKU) is approximately $3000-$10,000 dollars/year. However, the cost for caring for a poorly or untreated child with PKU or other metabolic disease and subsequent mental retardation can be tremendously expensive, especially if placement in a group home facility is required.

The North Dakota Chapter of the American Academy of Pediatrics have been very pleased with the laws that were passed in 1997 and 1999 regarding treatment for these metabolic diseases. We strongly support Senate bill #2239 and feel it strengthens the screening for metabolic diseases and very importantly the treatment for these metabolic diseases thereby assuring that no child in North Dakota will needlessly develop mental retardation because of inability to maintain an appropriate diet.

I urge you to help assure that these unfortunate children continue to be diagnosed and treated appropriately and protected from the ravages of mental retardation. This will provide protection for our most precious resource, the children of North Dakota. I would strongly urge you to pass the Senate bill #2239.

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 or concerns, 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!

SB 2239

Testimony on SB 2239
Regarding the Newborn Screening and Metabolic Food Programs
before the
Senate Appropriations Committee
by
Darleen Bartz, Department of Health
February 9, 2001

Good Morning Senator Nething and members of the Committee, I am Darleen Bartz Chief of the Preventive Health Section of the state Department of Health. With me is Karen Oby, Newborn Screening Program Coordinator, for the Maternal and Child Health Division. I am pleased to present this testimony regarding SB 2239 which revises the newborn screening law and clarifies the Department of Health's medical food program and client eligibility for coverage of benefits.

In 1967, the legislature enacted Chapter 25-17 which established the testing program for phenylketonuria (PKU) and other metabolic diseases. Under this law, the Department was directed to make arrangements for necessary treatment if a family was unable to pay. For many years, the Department has provided medical food to all families without regard to income.

In 1997, a new section was added to Title 26.1 (North Dakota Insurance Code) which imposed a mandate on all health insurance companies to provide coverage for treatment. In 1999, this mandate was modified to make the Department the primary payer. The Department was to commence a rulemaking process following this session to establish income eligibility for benefits. In May 1999 and again in March 2000 the Department published proposed rule. Due to opposition, the Health Council accepted the Department's recommendation to suspend rulemaking and develop legislation that would clarify eligibility for benefits under the metabolic food program.

A workgroup, composed of 20 individuals was developed to examine this issue. This group included legislators with affected families in their districts, representation from the ND Medical Association including four physicians, consumer representation, an individual from the Governor's Office, Health Department staff, and an individual from the Legislative Council. The consensus of the workgroup was that:1) PKU and Maple Syrup Urine Disease (MSUD) are significant public health issues and that medical food should be provided regardless of income; 2) insurance should be the first payer; 3) there should be a mechanism for the Health Department to collect from insurance companies, and others, and the funds go into the operating expenses for the Department; 4) there should be a provision by which individuals who are not eligible for benefits can buy metabolic food through the Department; and 5) the Department would need additional funding to pay for this funding this treatment program. These concepts were incorporated in the first copy of this bill. As a result of the initial hearing on this bill, amendments were proposed and adopted which again made the Department the primary payer and eliminated the option for the Department to seek payment from insurance companies.

In general, the engrossed version of SB 2239 amends the newborn screening and metabolic food program to permit the Department of Health to make changes in the newborn screening program that incorporate advances in the technology of testing for metabolic diseases and the recommendations of the American Academy of Pediatrics. The bill also gives explicit rulemaking authority to the Department of Health. Third, the amendments modify the law to reflect current practices and define more accurately the division of responsibility between Department of Health and Department Human Services for short-term and long-term follow up of children with special health care needs.

Due to the increased costs of medical food provided to individuals with PKU and MSUD and the increased program participation, it is projected that $80,000 will be needed during the coming biennium to continue to cover the treatment costs for all individuals with PKU and MSUD. If this bill is passed, the Department will need $80,000 from the general fund.

This concludes my prepared testimony. Either Karen or myself would be happy to respond to any questions you may have.

Thank you.

Estimated Costs for MCH Program for FY '02-03

Client	2000 (Actual)	FY '02 (Est)	FY ' 03 (Est)	
1	$ 3,669	$ 3,669	$ 3,669	
2	$ 3,669	$ 3,669	$ 3,669	
3	$ 3,669	$ 3,669	$ 3,669	
4	$ 3,792	$ 3,792	$ 3,792	
5	$ 1,039	$ 4,160	$ 4,160	Working to get back on diet
6	$ 1,773	$ 4,160	$ 4,160	Working to get back on diet
7	$ 3,792	$ 3,792	$ 3,792	
8	$ 4,986	$ 4,984	$ 4,984	
9	$ 1,930	$ 4,160	$ 4,160	Adult back on diet last part of 2000; anticipate full participation FY '02-03
10	$ 2,078	$ 4,160	$ 4,160	Getting back on diet, anticipate full participation in FY '02-03
11	$ 4,122	$ 4,122	$ 4,122	
12	$ 3,160	$ 3,160	$ 3,160	
13	$ 594	$ 4,160	$ 4,160	Adult moved to ND, getting back on diet in 2000, anticipate full participation in future)
14	$ 6,077	$ 6,077	$ 6,077	
15	$ 1,532	$ 4,160	$ 4,160	Trying to get young woman back on diet
16	$ 750	$ 4,160	$ 4,160	Trying to get young woman back on diet
17	$ 3,476	$ 3,476	$ 3,476	
18	$ 5,481	$ 5,481	$ 5,481	
19	$	$	$ 4,160	Comes off WIC in 2003
20	$ 3,792	$ 3,792	$ 3,792	
21	$ 4,688	$ 4,688	$ 4,688	
Total	$ 64,069	$ 83,491	$ 87,651	

	FY '02 (Est)	FY ' 03 (Est)	
New PKU	$ 2,950	$ 2,950	New PKU baby in FY '02
New PKU		$ 2,950	New PKU baby in FY '03
New Formula	$ 6,000	$ 6,000	Changes of current clients to more expensive diet
Low Pro	$ 4,000	$ 4,000	Low-pro foods for Medicaid Clients
Total	$ 96,441	$ 103,551	

Biennium Total	$ 199,992
Minus MCH	$ 120,000
Gen Funds	$ 79,992

532236

01-29-01

Good morning Madam Chairman, members of the House Human Services Committee, my name is Brenda Rakoczy, I am the mother of Sydney who is 5 and has PKU. Sydney is unable to make it today due to some conflicting appointments, but as you can see by this national calendar, which she was selected for the cover, she is one of the many PKU success stories you see in this room. I want to thank you for the honor of testifying in front of the "People's Legislature" **in favor of SB 2239.**

PKU is a rare metabolic disorder that causes profound mental retardation through the build-up of phenylalanine (PHE) in the brain and nervous system. If treatment begins shortly after birth, these affects can be avoided. As a result of state-mandated newborn screening and early and continued treatment, people with PKU can go on to lead normal lives and attain many of their goals and aspirations. PKU newborn screening has become compulsory since 1963 in the United States. There has not been a child born with PKU in North Dakota in the last three years.

Without SB2239, several barriers **(mostly financial)** will remain in preventing and treating this disease among our state's children and young adults. Quite simply, the treatment of PKU is one of the most economically valid and morally responsible investments we can make in our society. Thus the need for special metabolic formulas (medical food), low protein modified food and a way to pay for it – will remain throughout a PKU individual's life.

If individuals with PKU are not able to maintain a strict dietary regimen for life they become victims of a slow, but quite significant intellectual and neurological deterioration over a period of time. Following a very restricted low protein diet can prevent this terrible development; this regimented diet restricts all meats, dairy, poultry, legumes, nuts, and most products containing flour and grains. A typical diet includes prescription metabolic formula, small amounts of preciously weighed fruits and vegetables naturally low in protein and expensive low protein modified foods.

Doctor Levy, Children's Hospital – Boston, stated: "Generally as a result of such a strict diet so low in phenylalanine, so low in protein that a person without PKU on this diet would go blind or either die. But it doesn't necessarily mean what most of us consider a healthy diet of fruits and vegetables and salads. These also contain phenylalanine. People with PKU vary in their ability to process phenylalanine (PHE). Some can get away with eating salads or even pizza with no cheese or meat. For others, however, four lettuce leaves, or an ear of corn is an entire day's quota of PHE." (As very similar to my daughter's Sydney's allowance – of only 275 mg per day), as she grows her tolerance does not necessarily increase it can stay the same even when she is 40. We think of everything that goes into her mouth as medicine, and it is a constant challenge to meet her hunger and nutritional needs without adding the harmful phenylalanine, which would have a direct toxic affect on her body.

The heart of the problem now encountered by a family whose child is diagnosed with PKU is no insurance or lack of appropriate coverage (due to self funded plans and federal insurance plans) and possibly severe financial cuts to the formula program from the Health Dept if this bill is not approved. On behalf of the PKU families we would like it noted on legislative record that the true legislative intent of SB 2374 which was heard last legislative session was to have a combined cap for both medical food (formula) and low protein modified foods at $3,000 total. While it would have been great to have no fiscal limit, however, we feel it is imperative to be honorable and have the capped instituted as was originally testified to during the last two legislative sessions.

As a PKU individual or child grows so will their intake of formula. Sydney's prescription metabolic formula cost is currently $152.00 for 2 cans, she goes through approximately 8 cans per month at $608.00, than the metabolic flavoring to disguise the terrible taste of the formula is $46.00 for 6 cans, she uses approximate 5 cans per month. Low protein modified foods run approximately $50 - 100 per month. Keep in mind she is only five years old. Sydney prescription medical food not including her low protein modified food is over $7,296 per year. I'm honorably retired from the Air Force and Tricare is a federal insurance company and does not cover the prescription medical formula or low protein modified foods. Sydney's monthly medicine expense is higher than our home mortgage.

Without assistance from the state health department, we would have a very difficult financial barrier to providing for her immediate critical medical needs. Without passage of this bill one of these ND children will fall through the cracks.

Off-diet PKU/MSUD individuals are literally lives in danger. Stopping the diet can cause mental retardation, learning disabilities, behavior problems, neurological problems, personality disorders including schizophrenia, panic attacks and agoraphobia. The cost of metabolic formulas are minuscule compared to the cost of housing and educating a child with mental retardation which is approximately $100 – 125,000 per year.

I would like to personally thank the Health Dept for providing the medical formula for the families over the last 30 years. North Dakota is a forerunner in treating and preventing mental retardations in our state's most precious resources – our children. Additionally, I would like the thank the insurance companies for covering up to $3,000 dollar for low protein modified foods for individuals with true insurance coverage and for those individuals who are over the age guidelines for benefits from the Health Dept. Combining our efforts – we can ensure no individual falls through the cracks. It is too high a price to pay.

During the 56th Legislative Assembly - the Health Dept testified and we PKU families strongly concur with their position that the purpose of the Dept's Program – is to avoid the cost and burden of severe mental retardation.

"The Department will be able to maintain a register and assure that all eligible individuals, particularly children, receive necessary formula and low protein modified food products. If insurance companies had this responsibility, those with metabolic conditions could be subject to high deductibles or co-payments, and the Dept would not have the 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."

In further testimony they stated, "individual would move on and off health insurance coverage or change policies under which they have different co-payments and deductibles that could disrupt the continuity of these special dietary supplements."

As stated in their testimony the Health Dept is in the best position to ensure individuals' get their formula without any interruptions. The state of ND 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 Dept of Health has provided formula that have helped many families and children over the years."

It should be of special note that the Health Dept has provides formula for families who do not qualify for Medicaid. Individuals who qualify for Medicaid receive their formula through that program and some low protein modified foods from the Health Dept Maternal Childhood (MCH) Block Grant. The rest of the families who get formula through the Health Dept do not get low protein modified foods from the Health Dept they are appropriated through personal income or through an individuals insurance company if they have appropriate coverage.

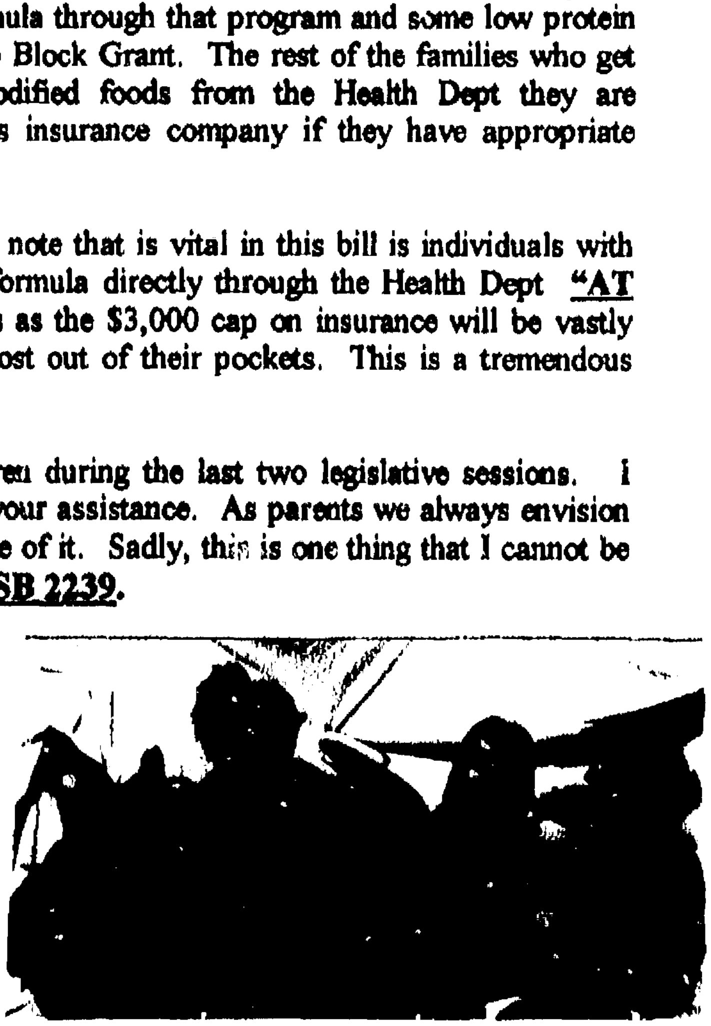
We PKU families certainly support this position. One additional note that is vital in this bill is individuals with PKU/MSUD over the age limits be authorized to purchase the formula directly through the Health Dept "AT COST." This will save astronomical mark-ups from pharmacies as the $3,000 cap on insurance will be vastly eaten up and those without insurance will have to pay the full cost out of their pockets. This is a tremendous attribute of this bill.

In closing, I thank you for your commitment to our PKU children during the last two legislative sessions. I would like to express how humble I am to have to come ask for your assistance. As parents we always envision when our children have a need, we will always be able to take care of it. Sadly, this is one thing that I cannot be my child's "all and all" – I need your help. **Please vote yes on SB 2239.**

Very Respectfully

Brenda J. Rakoczy

Brenda and Tom Rakoczy
1216 – 2nd Ave NE,
Reynolds, ND 58275
701-847-2047 Email: BRENDARAKOCZY@AOL.COM



March 2, 2001

To Whom It May Concern:

I am the mother of a beautiful, healthy 4 1/2 year old daughter named Kaylin Jo who has a metabolic disorder which is known as PKU.

Kaylin was officially diagnosed at 7 days old after a simple blood screening was done at birth. This simple test, which the state of North Dakota requires, may have saved her from a life of severe disability and saved taxpayers thousands of dollars. Kaylin was immediately prescribed a metabolic formula from a local pediatrician in our area, Dr. Lunn. She was also referred to a specialist in Fargo, ND whom had extensive knowledge in metabolic disorders, Dr. Kenien.

Kaylin began as an infant drinking the metabolic formula and has done very well with it. She, even to this day, loves her "milk". My husband and I have been fortunate in that we have not had problems getting her to drink her formula and therefore she has grown "like a weed". She currently is in the 95% for growth which in my eyes must mean we are doing something right.

As you may or may not know the formula is a staple in PKU children & adults diets. We have been told that this formula is extremely costly if we were to purchase it ourselves and are so thankful that the state pays for and is able to obtain the formula for us.

My husband and I both work out of the home full-time and still struggle as many families to do to make ends meet. We have 3 small children therefore have many additional household expenses including daycare. If we were required to purchase the formula ourselves I fear many of my children's other needs may go unmet.

Kaylin is fortunate in the fact that her body seems to be able to process a substantial amount of phe per day. We have been told that her formula may contribute to this. She typically requires about 600 mg per day but has been given up to 800 mg per day with good blood test results. My husband and I are also fortunate because this means we don't have to purchase as much low protein foods as other PKU families do. Kaylin is able to eat "normal" food which is low in protein and occasionally some specialty low protein food.

Senate Bill 2239 is important to our family because even with our health insurance coverage we would have a large out-of-pocket expense for formula in excess of what our health insurance would cover plus approximately $500 a year in low protein foods that must be purchased and shipped to our home.

Please vote "yes" on this Bill.

Thank you,

Tammy J. Anderson
Mother to Kaylin Anderson
Manvel, ND

Testimony in Support of SB 2239
House Human Services Committee
March 12, 2001
Robert M. Wentz, M.D., M.P.H.

I wish to indicate my support for SB 2239 regarding provision of special formula and foods for persons with PKU and other metabolic disorders.

Having spent 16 years of my career with the State Department of Health, including 8 years as Health Officer, I am well aware of the metabolic testing and treatment program. Since its inception, the program has provided testing for thousands of newborn infants in North Dakota and special formula and foods for many children with PKU. These services have allowed children with PKU to grow up to be healthy and productive adults and need to be continued.

I have personal experience with metabolic disorders as I have one called Fabry disease which has resulted in frequent episodes of pain, renal failure and heart rhythm problems requiring placement of a heart pacemaker. Metabolic disorders are genetic diseases and occur in many different forms. Treatments are being developed for more metabolic diseases every year and the Human Genome Project will accelerate these advances.

As a pediatrician with experience with PKU and a person with a metabolic disorder I urge a "Do Pass" on SB 2239.

Thank you.

Testimony on SB 2239
Regarding the Newborn Screening and Metabolic Food Programs
before the
House Human Services Committee
by
Darleen Bartz, Department of Health
March 12, 2001

Good Morning Chairman Price and members of the Committee, I am Darleen Bartz, Chief of the Preventive Health Section of the State Department of Health. I am pleased to present this testimony regarding SB 2239 which revises the newborn screening law and clarifies the Department of Health's medical food program and client eligibility for coverage of benefits.

In 1967, the legislature enacted Chapter 25-17 that established the testing program for phenylketonuria (PKU) and other metabolic diseases. Under this law, the Department was directed to make arrangements for necessary treatment if a family was unable to pay. For many years, the Department has provided medical food to all families without regard to income.

In 1997, a new section was added to Title 26.1 (North Dakota Insurance Code) which imposed a mandate on all health insurance companies to provide coverage for treatment. In 1999, this mandate was modified to make the Department the primary payer. The Department was to commence a rulemaking process following this session to establish income eligibility for benefits. In May 1999 and again in March 2000 the Department published proposed rule. Due to opposition, the Health Council accepted the Department's recommendation to suspend rulemaking and develop legislation that would clarify eligibility for benefits under the metabolic food program.

A workgroup, composed of 20 individuals was developed to examine this issue. This group included legislators with affected families in their districts, representation from the ND Medical Association including four physicians, consumer representation, an individual from the Governor's Office, Department of Health staff, and an individual from Legislative Council. The consensus of the workgroup was that: 1) PKU and Maple Syrup Urine Disease (MSUD) are significant public health issues and that medical food should be provided regardless of income; 2) insurance should be the first payer; 3) there should be a mechanism for the Department of Health to collect from insurance companies and others, with the funds going into the operating expenses of the Department; 4) there should be a provision by which individuals who are not eligible for benefits can buy metabolic food through the Department; and 5) the Department would need additional funding to pay for this treatment program. These concepts were incorporated in the first copy of this bill. As a result of the initial hearing on this bill, amendments were proposed and adopted which again made the Department the primary payer and eliminated the option for the Department to seek payment from insurance companies.

In general, engrossed versions of SB 2239 amend the newborn screening and metabolic food program to permit the Department of Health to make changes in the newborn screening program that incorporate advances in the technology of testing for metabolic diseases and the recommendations of the American Academy of Pediatrics. The bill also gives explicit rulemaking authority to the Department of Health. Third, the amendments modify the law to reflect current practices and define more accurately the division of responsibility between the Department of Health and the Department of Human Services for short-term and long-term follow up of children with special health care needs. Last, the Department may seek reimbursement from other government programs which provide coverage.

Due to the increased costs of medical food provided to individuals with PKU and MSUD and the increased program participation, it is projected that $80,000 will be needed during the coming biennium to continue to cover the treatment costs for all individuals with PKU and MSUD. The appropriation for this bill is included in SB 2004.

This concludes my prepared testimony. I would be happy to respond to any questions you may have.

Thank you.

Testimony for SB 2239
House Human Services Committee
March 12, 2001

Madam Chair and committee members, for the record I represent Blue Cross Blue Shield of North Dakota. This bill will hopefully correct a problem resulting in a bill approved by the 1999 Legislature. That bill established a $3,000 insurance mandate cap. The intent of that bill was that the formula (medical food) was to be covered by the Health Department and those with insurance could use their insurance coverage for the low protein modified foods. However, by mistake, the way the bill was interpreted that there was only a $3,000 limit on low protein modified foods and no limit on the formula. This was not the understanding and agreement reached with the parents of children with a metabolic disease and BCBSND.

During the interim, the bill sponsors worked to develop a revised bill, which would provide free formula and still a $3,000 cap that could be used for the low protein modified food. The parents were very supportive and good to work with during this process. The essence of the bill states that the Health Department will provide formula free to men up to the age of 22 and women up to the age of 45. In addition, parents can purchase formula from the department at cost in case there is a need beyond the established age deadlines. Those will insurance can use their $3,000 insurance limit for this additional formula need or for low protein modified foods. This gives the parents flexibility on how they want to use their insurance without penalizing them.

Madam Chair and committee members, BCBSND supports SB 2239 as amended in the Senate and urge you to give SB 2239 a Do Pass.

Rod St. Aubyn
Government Relations
Blue Cross Blue Shield of North Dakota

Bill #2239
Nancy Barrett Ouderkirk and Evan Ouderkirk
3-12-01

My name is Nancy Ouderkirk and this is my 5 year-old son, Evan. It has been five years since we first stood before you on behalf of North Dakota's PKU and MSUD children. And even though you may not remember us individually, you've had the opportunity to witness a success story.
When we were here initially, Evan was an infant. A great deal of uncertainty laid ahead for him. However, during these last 5 years, not only I have learned to better handle his disorder but so has Evan. Evan helps to make his own formula most mornings. He counts 17 tablespoons as I pour them into the blender and he mixes it. He helps mix up his own pancakes. Evan also rolls out his own tortillas and chips. Evan gets his own snack for school and puts it in his backpack. He even teaches me to play some of his computer games. Aside from Buzz Lightyear being one of Evan's favorite TV shows, he also likes to watch the History Channel. On numerous occasions Evan has enjoyed telling anyone who will listen about the Battle of Williamsburg and Haunted History. Evan is in preschool. He also is in the preschool choir at our church. He picks on his sister and plays with his friends.

Evan may not be seen as a remarkable child. He may seem like any other normal 5 year-old boy. Thank God! And thank God that the majority of other children don't have to go through what these kids have to go through to remain normal. These young ladies and gentlemen are all success stories. And for this reason I wish with deepest sincerity to thank all the professionals who have worked with our children, all of you, the Health Department, the Insurance companies, and the parents. It's because each one of these entities has worked in the best interest of PKU and MSUD children that we're all success stories. Because of all of us none of these children have fallen through the cracks or worse, had to be institutionalized. It is our hope that the current assistance we receive from the state will continue. With that having been said, please don't think that we are expecting the entire concern for our children's health be shouldered by the Health Department. We as parents very definitely do our part.

Aside from the formula, the majority of Evan's dietary needs are met by what I can purchase from the grocery store. However, being Evan can't eat any meat products, dairy products or any high protein food items, this also means he can't eat anything containing flour, eggs, soy or nutrasweet. As you may have noticed this greatly limits what he can eat. Therefore, it is necessary that I special order additional foods for him.

I recently ordered 6 food items for Evan – 4 boxes of pasta, one box of rice, and one box of chocolate flavored candy. The bill came to $50.30. That averages @ $8 per box. Out of necessity Evan is a very strict vegetarian. He eats low protein pasta; low pro breads and we make our own pizza crust. In addition to daily monitoring of Evan's food intake, there is the preparation of the food. I'm sure that when you are at home preparing your meals, most of you only prepare one meal per sitting. However in our house, at most meal times I must prepare two main courses. If we are having macaroni and cheese, I prepare Evan's special ordered cheese and his pasta. And then I make regular macaroni and cheese for my daughter and myself. Doing this for one day is no big deal. But continuing this day after day requires a great deal of organization and forethought. Even though Evan can eat fries from Mc Donald's, there's @ 128 mgs. of phe per small serving. That's over half of his daily diet and doesn't usually fill him up. And he can no longer eat their salads. Consequently, we don't go there often. The closest thing to fast food Evan really has is what I make ahead and freeze. Going on a family vacation means that we take it all with us because we most likely can't buy it when we get there.

Even something as simple and fun as Halloween takes more thinking ahead than the average family. We want Evan to be as much like any other child as possible. We don't want PKU to ever be a reason for him to not participate. That's one reason why every year at Halloween I prepackage candy items and I deliver them to each of the houses in our cul-de-sac so that when Evan rings their door bell he will receive something just like every other child who goes to that door.

Evan's current dietary needs cost approximately $4700 per year. Each case of formula costs the Health Department about $160. Evan uses @ 2 cases per month which is $320 per month - $3840 per year. His special ordered low protein food costs are approximately $70 per month - $840 per year.

We strongly hope you will support Bill #2239 as it represents a guaranteed program. It's a guarantee that if these children are provided with their formula (and ingest it) that they will remain mentally healthy. The other side of the coin is...that if these same children do not receive their formula, it is guaranteed that they will experience mental retardation and the cost emotionally and financially for the parents, children and the Health Department will be much greater. By voting in support of Bill #2239, you will guarantee continued success for the PKU and MSUD children of the state of North Dakota. On behalf of Evan, his sister Kailynn, their father and myself we thank you for your consideration.

Good Morning!

My name is Kris Wangler. My husband, Dale, and I have been blessed with six wonderful children and we farm 10 miles southwest of Rugby, North Dakota. Our youngest child, Rebekah, who is now three and a half years old, was born with the inherited metabolic disorder of Phenylketonuria (PKU). Currently, I believe that Rebekah is the youngest child in the state to test positive for PKU. Thanks to the mandatory PKU testing of all newborn babies in our state and wonderful doctors and dieticians who specialize in metabolic diseases, Rebekah is a normal, bright, young girl today. The road ahead for her, unfortunately, is only a one way street, with no detours. In order to remain healthy and normal, Rebekah must be restricted to a very limited diet, which consists of only a few fruits, vegetables, and low protein modified foods. More importantly, she must drink a special formula that supplies her with the essential nutrition needed to grow and develop for the rest of her life. If Rebekah was to take a detour from this diet or stop taking her formula, the increased phenylalanine in her system would poison her brain, thus she would become mentally handicapped. I cannot begin to express to you the importance of the formula in the lives of PKU children. They need this formula like fish need water or plants need sunlight.

Because of the high costs of the formula and low protein modified foods, many of us parents cannot financially afford to purchase them for our children. We, the parents, and our children, have come here today to ask for your help in supporting Senate Bill #2239 in which the North Dakota State Department of Health would help supply formula to female children through the childbearing years of 44 and also mandate insurance companies to allow a cap of $3,000 for low protein modified foods.

Our beautiful state of North Dakota has taken the first step in helping children born with PKU and Maple Sugar Disease (MSD) by making blood test screenings mandatory on all newborns. It is now our prayer that you will help us to help them lead normal, healthy lives and take the next step in keeping these children in our state and out of institutions by supporting Senate Bill #2239.

God Bless You All!

Kristin M. Wangler

Chairman, and members of the Human Services Committee.

My name is David Aakre, I live in Kindred, ND with my four children, Danielle, DeAnna, Dolan, and Dalton. Danielle, Dolan, and Dalton are diagnosed with, and are being successfully treated for the metabolic disease called Phenylketonuria or PKU. DeAnna does not have PKU. Looking at them you can not tell who is diagnosed with PKU, and who isn't. If however the children with PKU were not treated you would have no problem telling who they were. They would likely be in wheelchairs, unable to control their actions, and incapable of engaging in normal conversation. From birth this disease silently, and slowly renders the individual with PKU to a totally dependent state of care and support if left untreated. Denying nothing as a man, this is an emotional issue. When a baby reaches two weeks of age, you start getting glimpses of the little personalities inside them. This is also the time notification is made that a baby has this metabolic disease. Without the proper treatment for this disease, at two months of age this personality will start to vanish. The rewards are immense when you make comparisons between a successful treatment program, and one that is not. There are many lessons in life that can change pride to thankfulness, and this is definitely one of them.

This is my fourth time here at the State Capital to take part in the legislative process, and it's still an awesome place. I can only imagine what it's like to be here on a daily basis, and what the feelings were when each of you first took office. There are some similarities between what you experience here, and what we as parents of the children with these metabolic diseases experience. The same feelings you have when you introduce new legislation, and have success in seeing it through the legislative process is just like us. These are milestones in your career, and is how your effectiveness is measured. You see when our children reach milestones in their lives such as smiling, rolling over, sitting up, walking, talking, starting school, participating in school activities, graduating, and leading productive lives, is where we measure our effectiveness in the treatment of these diseases. When each of these milestones are reached there is plenty of rejoicing, and at the same time apprehension for the next milestone.

This bill guarantees the continued success for the treatment of these metabolic diseases, and will ensure these children, and adults will have a chance to succeed in life. The consequences of having just one child not treated for this disease would be heartbreaking, and costly. This was the experience when the Guthrie test was first instituted. Children born just a few months prior to this were not treated, and consequently suffered irreversible neurological damage. The cost induced for the care of one individual not treated for this disease is much greater than the cost required to treat all the children, and adults in North Dakota. This is the testimony for the successful treatment of these metabolic diseases, which this bill supports.

Danielle recently has competed at speech events in the inform category, and chose PKU as her topic. She placed fifth her first time participating at a speech event at the Valley City Optimist speech tournament, and is well on her way to lettering her first year. For these children and adults to have the ability to succeed is something we can never take for granted. This bill, and the treatment program it supports will provide them that ability, and it is one we are so thankful for.

Thank you very much for your prayerful consideration for this legislation.

David

David C. Aakre
P.O. Box 84
Kindred, ND 58051

Testimony on SB 2239
Regarding the Newborn Screening and Metabolic Food Programs
before the
House Appropriations Committee
by
Darleen Bartz, Department of Health
March 20, 2001

Good Morning Chairman Timm and members of the Committee, I am Darleen Bartz, Chief of the Preventive Health Section of the state Department of Health. I am pleased to present this testimony regarding SB 2239 which revises the newborn screening law and clarifies the Department of Health's medical food program and client eligibility for coverage of benefits.

The engrossed version of SB 2239 amends the newborn screening and metabolic food program to permit the Department of Health to make changes in the newborn screening program that incorporate advances in the technology of testing for metabolic diseases and the recommendations of the American Academy of Pediatrics. The bill also gives explicit rulemaking authority to the Department of Health. Third, the amendments modify the law to reflect current practices and define more accurately the division of responsibility between the Department of Health and Department Human Services for short-term and long-term follow up of children with special health care needs. Last, the department may seek reimbursement from other government programs that provide coverage.

Due to the increased costs of medical food provided to individuals with PKU and MSUD and the increased program participation, it is projected that $80,000 will be needed during the coming biennium to continue to cover the treatment costs for all individuals with PKU and MSUD. The appropriation for this bill is included in SB 2004.

This concludes my prepared testimony. I would be happy to respond to any questions you may have.

Thank you.

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Title.0700

Prepared by the Legislative Council staff for
Representative Klinske
March 28, 2001

PROPOSED AMENDMENTS TO REENGROSSED SENATE BILL NO. 2239

Page 1, line 1, after "25-17" insert "and a new subsection to section 50-10-06"

Page 1, line 2, after "law" insert "and services for treatment of phenylketonuria and maple syrup urine disease"

Page 2, line 24, after "council" insert "and the department of human services"

Page 2, line 29, after the second period insert:

"1."

Page 3, line 1, overstrike "1." and insert immediately thereafter "a."

Page 3, line 4, overstrike "2." and insert immediately thereafter "b."

Page 3, remove lines 8 through 24

Page 3, line 25, replace "7" with "c"

Page 3, after line 26, insert:

"2. The department of human services, as a program provided under chapter 50-10, shall:

a. Provide medical food at no cost to males under age twenty-two and females under age forty-five who are diagnosed with phenylketonuria or maple syrup urine disease, regardless of income. If treatment services under this subsection are provided to an individual by the department, the department may seek reimbursement from any government program that provides coverage to that individual for the treatment services provided by the department.

b. Offer for sale at cost medical food to females age forty-five and over and to males age twenty-two and over who are diagnosed with phenylketonuria or maple syrup urine disease, regardless of income. These individuals are responsible for payment to the department for the cost of medical food.

c. Provide low-protein modified food products, if medically necessary as determined by a qualified health care provider, to females under age forty-five and males under age twenty-two who are receiving medical assistance and are diagnosed with phenylketonuria or maple syrup urine disease."

Page 5, line 12, after "a" insert "state", after "health" insert "or department of human services", and after the period insert:

"**SECTION 8.** A new subsection to section 50-10-06 of the North Dakota Century Code is created and enacted as follows:

<u>Provide medical food and low-protein modified food products under chapter 25-17 to individuals with phenylketonuria or maple syrup urine disease.</u>"

Renumber accordingly