Approved 3/12/84
Date

MINUTES OF THE HOUSE COMMITTEE ON PUBLIC HEALTH AND WELFARE.

The meeting was called to order by Marvin Littlejohn at
Chairperson

1:30 a.m./p.m. on February 29, 1984 in room 423-S of the Capitol.

All members were present except:

Rep. Jesse Harder, excused

Committee staff present:

Emalene Correll, Research
Bill Wolff, Research
Norm Furse, Revisor
Sue Hill, Secy. to Committee

Conferees appearing before the committee:

Representative Homer Jarchow
Dr. Sechin Cho, Assoc. Professor Pediatrics & OBGYN, K.U. Med. Center, Wichita, Ks.
Dr. Virginia Tucker, Medical Dir.Crippled & Chronically ill Children's Program, Ks. Dept. of Health and Environment.
Evelyn Smith, R.N., Ks. State Nurses' Association
Lila Paslay, Ks. Assoc. for Retarded Citizens, Topeka
Joan Strickler, Exec. Dir. Ks. Advocacy & Protective Serv.for Developmentally Disabled, Inc.
Frank Gentry, Kansas Hospital Association
Joan Remmers, Kansas Association Home Health Agencies
Diane Bottorff, Kansas Nurses' Association
Ken Schafermeyer, Kansas Pharmacists' Association

Visitor's register, (See Attachment No. 1.)

Chairman called meeting to order stating there was a failed attempt this date on the floor of the House to amend HB 3047 into HB 2796, and since there will be no time for hearings before the deadline facing committee on House Bills this week, what is the pleasure of committee in regard to taking action on HB 3047 without hearings.? There was some discussion of an amendment, so Chair set HB 3047 aside for a time, and committee will get back to this bill later today if there is time.

HB 2864 Hearings began:--

Representative Homer Jarchow spoke to HB 2864, giving a brief background on the testing for Galactosemia in newborn babies. He distributed a hand-out to committee, (see Attachment No. 2.), for details. The enzyme deficiency test we are asking for here he said is easily detected, and it can do much to stop mental retardation in many cases, and other related diseases to this enzyme deficiency. He has personal feelings for this bill and urged committee for their support of HB 2864. He then introduced Dr. Sechin Cho to speak to this issue and to speak specifically to line 50, and the number of days mentioned.

Dr. Sechin Cho, Medical Center in Wichita spoke very comprehensively about how this deficiency test will detect several problems in newborn infants. Also how very simply can be rectified by eliminating all milk and milk products from the child, and said the number of days mentioned in line 50 would not really present a problem. Further, he spoke to the problems of cataracts developing, low blood sugar, mental retardation. Liver damage can also occur, tho it is reversible. He spoke to the percentages where Galactosemia occurs, and that the testing, tho a separate test from PKU and Hypothyroid tests, can be done with the same droplet of blood taken by a heel prick from the infant. Commented that a soy formula is a good substitute for milk for the infants. He said that pre-natal testing can be done too, and is when a second child of the same two biological parents is expected after their first child showed Galactosemia when tested. By the 18th week of the pregnancy they are aware of a problem if one indeed exists, and milk products are withdrawn from the expectant mother. He answered questions from committee, and spoke in favor of this testing proposed in HB 2864.

MINUTES OF THE HOUSE COMMITTEE ON PUBLIC HEALTH AND WELFARE,

room 423-S, Statehouse, at 1:30 a.m./p.m. on February 29, 1984.

HB 2864 continues:--

Dr. Virginia Tucker, from Health and Environment gave hand-out, (see Attachment No. 3.), for details. She said, being a Pediatrician herself, she is indeed in favor of HB 2864. More and more information is becoming available all the time, and they can with improvement in techniques make diagnosis prior to the birth of the infant, as Dr. Cho spoke about. They can provide screening diagnostic controlled tests for newborns, maintain a registry of cases, provide the necessary treatment. She stated further, she feels in order for the screening tests to be done in a quality way, it will take some time to get set up for such screening. She spoke to the costs of the tests, saying if done on mass scale, they would cost only about $.50 per test, but if done individually, could run $3.50 or more. The costs to the state for the care of the mentally retarded child is up to $100.00 per day, so annually this is $36,000 cost per year, per child. It seems then, that if we could avoid this, she said, the cost of the testing seems quite small. There is no way to say what savings for child and families would be. This cannot be measured in dollars. She called attention to the hand-out, saying that it will show the outline of the disease, symptoms and etc. She urged for consideration of favorable passage of HB 2864. She stated further, the Department of Health and Environment recommends the bill for favorable passage as well. She then answered questions.

Chair then welcomed all the nurses in attendance of todays meeting, saying that this particular HB 2864 was scheduled for this date because of it's interest to those in the nursing profession.

Evelyn Smith, R.N., Ks. State Nurses Assoc. spoke to committee in support of screening for Galactosemia. Though it is a rare disease, it can be stopped. Because time is of the essence in these cases and milk creates grave problems for these babies, the screening tests may be livesaving procedures. She had written a paper published March, 1980 in the Nurse Practitioner, and it along with another journal article accompanies her testimony, (see Attachment No. 4.), for full details.

Lila Pasley, Ks. Assoc. for Retarded Citizens spoke as a volunteer and a parent of a mentally retarded child, saying they are extremely grateful to physicians, teachers, and other professionals, and to the state of Kansas. Our state has been among the leaders in applying technologies toward prevention of mental retardation, i.e. testing and diet for children with PKU and immunization against red measles and German measles. We support HB 2864, she said, as one more step in preventing one more cause of mental retardation.

Atch 5

Joan Strickler, KAPS, gave hand-out to committee of her testimony, (See Attachment No. 6.) for details. She said that we are so lucky that this method of screening can provide such a wealth of medical knowledge and said it makes good sense to use these techniques that are available to help. She urged committee for favorable support of HB 2864.

Mr. Frank Gentry, Ks. Hospital Association assured full cooperation from the Hospitals in this screening process, and says they support HB 2864.

Hearings closed on HB 2864.

3024 Hearings began:--

Joan Remmers, Kansas Association Home Health Agencies spoke to HB 3024. They would like to clear up some confusion about what the Home Health Agencies really does. They are Home Health providers that go into private homes to provide nursing care, physical therapy, occupational therapy, speech therapy, and home health aide services. This is done through contact from a physician. The physician sets up a plan of treatment, and they as the provider go in and follow through with the treatment. The members in our Kansas Association are all certified members she stated. They are seeing an influx of home care providers though that are not certified, and this causes some concerns. Those concerns deal with quality of good care; a patient living alone may have problems with giving themselves the proper dosage of medication, i.e., insulin; nursing liability; reimbursement problems; etc. We do encourage family members and or support groups, she said, to help these persons living alone so that they may remain independent. Our nurses aides are not required to have certification. They have no previous training in administering medications, nor is this training required. We feel that in a nursing home or institutional setting that nurse aides are not allowed to administer medications, and

HB 3024 continues:--

so we question that those in home health settings should be allowed. There was some discussion during questioning of Ms. Remmers in regard to reimbursement from medicare, how pre-filling syringes are kept sterile, complicated dosages of insulin resulting in problems, sedimentation of insulin, etc. She stated at the end of her testimony that their association supports HB 3024.

Diane Bottorff, Kansas State Nurses Association gave hand-out to committee, (see Attachment No. 7.), for details. Their Association is in strong support of HB 3024 she said. Home Health Aides are not required by all home health agencies to have completed a nurse aide course, so some have had no formal training in the care of sick and disabled. Pre-filling of insulin syringes is at best risky when done by aides, she said, since they do not have the training. Insulin is a potentially dangerous drug. Too much or too little insulin in the syringe can lead to severe complications or could even be fatal. We feel, she said this issue of dealing with medications should be within the scope of those who are best qualified to do so.

Ken Schafermeyer, Executive Director of Ks. Pharmacists' Association spoke to HB 3024, in that he agreed with what testimony had been given by the nurses today, and the Kansas Pharmacists' Assoc. is in full support of HB 3024. He gave hand-out of his testimony to committee, (see Attachment No. 8.), for details.

Chairman noted that he had spoken earlier with Jerry Slaughter, of Ks. Medical Society, who is unable to present testimony in person, and Mr. Slaughter had stated the Association he represents is in favor of HB 3024.

Hearings closed on HB 3024.

Chair recognized Rep. Wagnon and HB 3047 was brought up for discussion again.

HB 3047

Rep. Wagnon noted the basics of this bill are with the child in need of care information systems and these are specified in new section 1, and new section 2 of HB 3047. She then moved to amend HB 3047 in this manner. To strike all of sec. 3, (starting on Page 3), sec.4. sec.5, and sec.6, down to line 192 on page 5, and to re-number sec. 7., and to amend the title slightly to take out the reference to the protection order. Motion was seconded by Rep. Williams. Discussion. Vote taken, motion carried.

Rep. Wagnon then moved that HB 3047 be passed as amended out of committee favorably, seconded by Rep. Williams. Motion carried.

HB 2098

This bill has been referred back from the floor of the House to committee and a copy of a proposed substitute bill having been prepared by Revisor was handed to committee, (see Attachment No. 9.), for details. Revisor, Norm Furse read the ambiguities in the bill, then reading from the draft, the new language for the proposed substitute bill. He touched on all the technical aspects and answered question from committee regarding same. A motion to accept this substitute bill, HB 2098 was made by Rep. Williams, seconded by Rep. Cribbs. Discussion. Vote taken, motion carried.

Motion by Rep. Friedeman to insert the word, "Kansas" in section 1, (b) before the word law. Seconded by Rep. Blumenthal, motion carried.

Rep. Branson then moved this substitute bill for HB 2098 be reported out of committee as amended favorably for passage. Seconded by Rep. Williams, motion carried.

Meeting adjourned 2:55 p.m.

Date: 2-29-84

GUEST REGISTER

HOUSE

PUBLIC HEALTH AND WELFARE

Please Print

NAME	ORGANIZATION	ADDRESS
~~Renee Stotler~~	SN/ Bethel College	North Newton
Carmen Goering	SN Bethel College	N. Newton, Kansas
Jan Baty	SN Bethel College	Newton, KS
Nancy Noble	SN Bethel College	N. Newton, KS
Karen Mace	SN Bethel College	N. Newton, Ks.
NORA ALONZO	ST. MARY College	Leav. Ks.
DOROTHY LADD	Osawatomie St. Hospital	Paola, Ks.
Marlene Smith	Miami County Hospital	Paola, Ks.
Grace Wenger	KSNA sponsored Bethel College SN	N. Newton, Ks
Donna Klassen	SN/ Bethel College	North Newton, KS
Karlene Basta	SN/Cloud County C.C.	Beloit, Kansas
Rita McMillan	SN Cloud County C.C	Beloit, Kansas
Carolee Neufeld	~~RN~~, KSNA	Hillsboro, Kansas
Jacque Robbins	SN Wichita State University	Wichita Kansas 67208
Sheri Donaldson	SN Wichita State University	Wichita KS.
Bernadette Kennedy	KU School of Nursing	Lenexa, Ks.
Kathy Patterson	KSNA	1275 W 41st #3 KCMO 64111
Virginia L. Tucker, M.D.	KDHE	Forbes, Topeka
Susan Bergh	NS Wichita State University	1717 N. Yosemite #10 Wichita Ks 67208
Keri Hewitt	NS wichita State University	3330 N. Oliver #70E wichita, KS. 67220

Attm. #1.
2-29-84

Date: 2-29-84

GUEST REGISTER

HOUSE

PUBLIC HEALTH AND WELFARE

NAME	ORGANIZATION	ADDRESS
Marilyn Chamberlin	KSNA	1506 Univ. Dr. Lawrence
Hazel Harned	KSNA	2708 Colleen, Dodge City,
Jane Mullen	KSNA	909 Allison Manhattan Ks
Elizabeth McCoy	KANS	4165 E Central #246 Wichita, KS
Debbie Lutz	WSU	Andover, KS
Diane Skochn	Nursing student	Box 321 Montezuma Ks 67867
Pat Thompson	Nursing Student	Box 396 Ashland, KS. 67831
Frank Lipsky	Ks Hospital Assn	Topeka
Ted Woodbury		Topeka, Ks
BARBARA HRENCHIR	~~UTE~~ WSU	2330 N OLIVER WICHITA, KS
Ruth Groves	KECZ	Topeka
Sandy Webb	WSU	3133 Eveningside Way, Topeka 66605
Mary Jane Breeden	WSU	3517 S. Knight Wichita Ks 67217
Martha S. McDowell	WSU - Nurse Clinician Student	2 Beech Rd Wichita Ks 67206
Connie R. Luty	WSU - NS Clinician Student	448 Hickory Ln, Valley Center 67147

Date: 2-29-84

GUEST REGISTER

HOUSE

PUBLIC HEALTH AND WELFARE

Please Print

NAME	ORGANIZATION	ADDRESS
Lila Pasley	La Leche League, Retired Citizen	Topeka
Carol Borger	K.S.N.A. Dist X	Augusta Ks 1613 Sacony 6-1416
Sue Steitz	KSNA Dist. #2	Roeland Park, Ks.
Joellen Greischar RN	KSNA Dist #2	KANSAS CITY KS 66103

Date: 2-28-84

GUEST REGISTER

HOUSE

PUBLIC HEALTH AND WELFARE

Please Print

NAME	ORGANIZATION	ADDRESS
Nancy Utech	Nursing Student / WSU	Wichita 2330 N. Oliver #703
Jane Darby	Nursing Student / WSU	843 S. Terrace Wichita
Diana Donovan	Nursing Student / WSU	Rt 1 Valley Center
ELIZABETH PLUMMER	RICE Co. Dist #1 Hosp	Lyons, Ks. 67554
Rita Deeds	Hospital Dist. #1 of Rice Co.	Lyons, KS. 67554
Tami Poland	Nursing Student / WSU	11616 W. 33rd N. Wichita, KS 67204
Colette Balding	Nursing student / WSU	3105 Sierra Pkwy #4 Hutchinson, Kansas 67502
Jane Davis	Dist IV KSNA	926 N. Spencer Newton, KS 67114
Maureen Wedel	Dist IV KSNA	823 Spruce Halstead, Ks 67056
Ann Ernst	Dist VI KSNA	Wichita Ks. 67203
LAURA Harper	Dist. VI KSNA	Wichita, Ks 67212
Pat Thompson	Dist III KSNA	Parsons, Ks 67357
Kim McMunn		Parsons, Ks 67357
Lisa Shaw	Dist VI KSNA WSU Nursing Students	316 Cardinal Ln. Wichita, Ks 67230
Lisa Hasdeallor	Dist VI KSNA WSU Nsg Student	1103 N Harding Wichita 67208
Garrie Standiferd	Washburn U. Nsg. Student	RR#2 Mayetta, Ks. 66509
Delaine Smith	W. U. Nsg. Student	1500 College, Topeka, KS. 66604
John Kelly	Topeka	Dept of Human Resources
Ann Filer	KSNA Dist 17	210 Yorkshire DR. Lawrence, Ks.
Bonnie Cather	Instructor - Labette Comm.	Parsons Ks
Leota McConnell	Instructor - College	Parsons Ks
#4 Gayla Todd	Nursing student	Greensburg <---> Dodge City Community College
Jeanne Schmidtt	Nursing student	Kinsley <---> Dodge City Community College

484.

Attm. # 2
2-29-84

HOMER E. JARCHOW
REPRESENTATIVE, NINETY-FIFTH DISTRICT
SEDGWICK COUNTY
2121 WEST DOUGLAS
WICHITA, KANSAS, 67213

STATE OF KANSAS

TOPEKA

HOUSE OF
REPRESENTATIVES

COMMITTEE ASSIGNMENTS
MEMBER: ASSESSMENT AND TAXATION
COMMERCIAL AND FINANCIAL
INSTITUTIONS

February 28, 1984

HOUSE BILL NO. 2864

I WOULD LIKE TO THANK THE CHAIRMAN FOR THIS HEARING ON HOUSE BILL 2864. MY DAUGHTER-IN-LAW ALSO THANKS YOU. IT WAS HER IDEA THAT I PURSUE THE ISSUE CONTAINED IN THE BILL.

HOUSE BILL NO. 2864 ADDS THE REQUIREMENT OF TESTING FOR GALACTOSEMIA ON NEW BORN BABIES. AS YOU CAN TELL BY THE BILL PKU AND HYPOTHYROID TESTS ARE PRESENTLY REQUIRED. I HAVE BEEN ADVISED THAT THE TEST FOR GALACTOSEMIA CAN BE ACCOMODATED FROM THE SAME DROPLET OF BLOOD TAKEN BY A HEEL PRICK THAT IS USED FOR THE PKU AND HYPOTHYROID TESTS.

I HAVE ATTACHED A COPY OF AN ARTICLE FROM THE JANUARY - FEBRUARY ISSUE OF THE SATURDAY EVENING POST WHICH OUTLINES THE GALACTOSEMIA PROBLEM AND CORRECTION IN WORDS MY DAUGHTER-IN-LAW COULD UNDERSTAND. SHE IS THE MOTHER OF THREE HEALTHY CHILDREN BUT HAS A SPECIAL PLACE IN HER HEART FOR RETARDED CHILDREN.

THE ENZYME DEFICIENCY TEST APPARENTLY IS EASILY DETECTED. GALACTOSE IS A MILK SUGAR THAT NEEDS AN ENZYME TO CONVERT IT INTO GLUCOSE; OTHERWISE THE GALACTOSE ACCUMULATES DANGEROUSLY IN THE BLOOD. THE CORRECTION IS TO TAKE THE BABY OFF MILK IMMEDIATELY OR, IDEALLY, NEVER BE STARTED ON MILK. IF MILK FEEDING IS CONTINUED, THE BABY WILL NOT ONLY BECOME MENTALLY RETARDED, BUT WILL DIE AT AN EARLY AGE.

Attm. #2
2-29-1984

DR. SECHIN CHO, WITH THE K.U. MEDICAL CENTER --- WICHITA BRANCH ADVISED ME THAT HE IS CURRENTLY ATTENDING TWO GALACTOSEMIA PATIENTS. YOU WILL BE HEARING FROM HIM AS A SPECIALIST AND PROPONENT OF THE TEST. DR. LEONA THEROU AND DR. NEIL SCHIMKE OF THE K.U. MEDICAL CENTER, ALTHOUGH NOT SURE THEY COULD ATTEND THIS HEARING, ALSO ARE IN FAVOR OF THE TESTING. DR. HOLLOWELL, THE DIRECTOR OF THE DIVISION OF HEALTH, ALSO IS IN FAVOR OF THE TESTS AND ADVISED ME HE WOULD HAVE SOMEONE HERE FOR THE HEARING.

I HAVE A CONCERN ABOUT THE NUMBER OF DAYS ON LINE 50 OF THE BILL. I WOULD APPRECIATE THE DOCTORS SPEAKING TO THE NUMBER OF DAYS FOR THE COMMITTEES CONSIDERATION.

THE ROUGH COST FIGURES OF THE GALACTOSEMIA TESTS THAT I HAVE BEEN FURNISHED ARE RELATIVELY MINOR. A ONE TIME NON-RECURRING COST OF $10,000 FOR EQUIPMENT AND RECURRING COSTS OF APPROXIMATELY $25,000 A YEAR (IF ALL TESTS WERE DONE IN ONE CENTRAL LAB). AS DR. KOCH SAYS IN THE POST ARTICLE "ALL YOU HAVE TO DO IS FIND ONE PATIENT, AND IT PAYS FOR ALL THE SCREENING".

ARE THERE ANY QUESTIONS?

Speaking Out

A MENTALLY RETARDED BABY IS FOREVER

Severe mental retardation and death from galactosemia can be prevented with screening and early treatment. If mothers passed the laws, all babies would be screened. You can help.

by Cory SerVaas, M.D.

State legislatures are important because they can pass laws that change people's lives.

I can't think of a more drastic change in a young woman's life than to become the mother of a mentally retarded child, when for a few cents a day, that child, if treated in the first days or weeks of life, could have developed a normal brain.

All but a few of our states have passed laws that require *all* babies to have a heel prick with a droplet of blood drawn to test whether the child may have hypothyroidism.

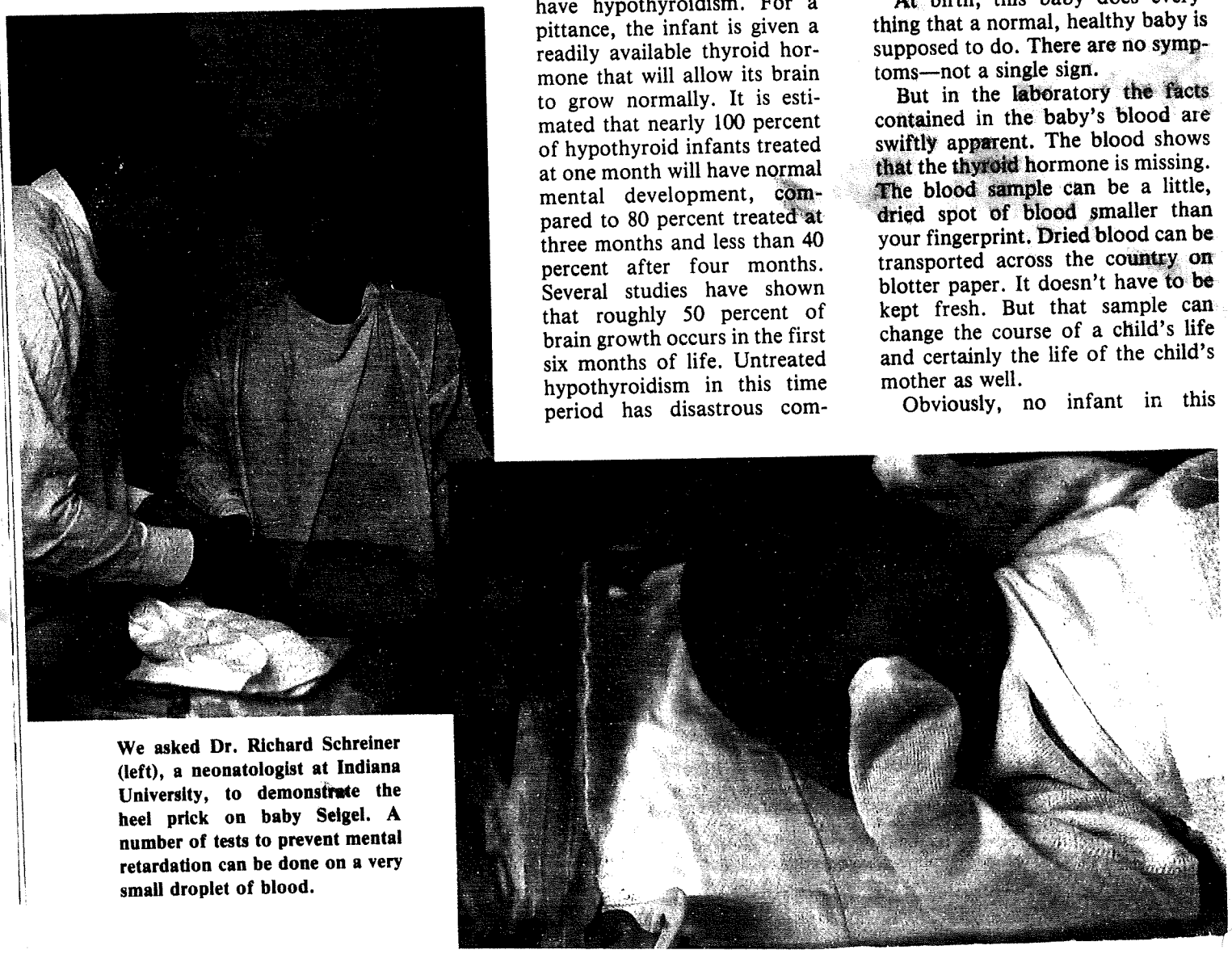
One out of 4,000 infants born in the United States is found to have hypothyroidism. For a pittance, the infant is given a readily available thyroid hormone that will allow its brain to grow normally. It is estimated that nearly 100 percent of hypothyroid infants treated at one month will have normal mental development, compared to 80 percent treated at three months and less than 40 percent after four months. Several studies have shown that roughly 50 percent of brain growth occurs in the first six months of life. Untreated hypothyroidism in this time period has disastrous complications in the baby's brain.

Yet there are no symptoms, and there is nothing about the pregnancy of the healthy mother who bears a hypothyroid baby to give even a clue that the baby will not develop normally. No doctor can tell by physical examination. The most experienced pediatrician cannot tell.

At birth, this baby does everything that a normal, healthy baby is supposed to do. There are no symptoms—not a single sign.

But in the laboratory the facts contained in the baby's blood are swiftly apparent. The blood shows that the thyroid hormone is missing. The blood sample can be a little, dried spot of blood smaller than your fingerprint. Dried blood can be transported across the country on blotter paper. It doesn't have to be kept fresh. But that sample can change the course of a child's life and certainly the life of the child's mother as well.

Obviously, no infant in this



We asked Dr. Richard Schreiner (left), a neonatologist at Indiana University, to demonstrate the heel prick on baby Seigel. A number of tests to prevent mental retardation can be done on a very small droplet of blood.

civilized country should be permitted to grow up mentally retarded from this highly preventable cause. A small amount of thyroid hormone replacement given orally every day allows the infant's brain to grow and function normally.

The only remaining states that do not have the mandatory testing law are Virginia, Mississippi, Nebraska and Hawaii. These states have voluntary programs for screening their newborns, but they are not mandatory. Virginia has passed a law requiring hypothyroid screening that will be in effect on July 1, 1984.

Another preventable cause of mental retardation is phenylketonuria (PKU). PKU is the inability of the infant to metabolize the amino acid phenyalanine, an amino acid present in protein that accumulates in the blood. By decreasing this amino acid in the diet during the infant's rapid-brain-growth period, mental retardation can be avoided. Testing for PKU is mandatory in every state except Mississippi.

Galactosemia

The test that The Saturday Evening Post Society wants to crusade for at this time is another enzyme deficiency test that can be easily detected—from the same drop of blood already mandatorily drawn from the baby for the PKU and hypothyroid tests. The next most common cause of preventable mental retardation is *galactosemia*, which simply means galactose in the blood. Galactose is a milk sugar that needs an enzyme to convert it into glucose; otherwise the galactose accumulates dangerously in the blood. If the baby doesn't have the enzyme, it will be fine as long as it isn't fed galactose. The only natural source of galactose is primarily in dairy products, so the baby should be taken off milk immediately or, ideally, never be started on milk. If milk feeding is continued, the baby will not only become mentally retarded, but will die at an early age.

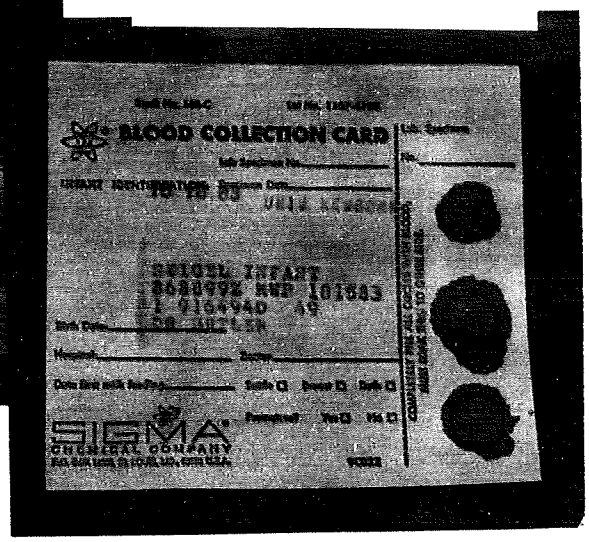
In one typical family the first child, a boy, died from what was presumed to be pneumonia at one month of age. Unfortunately, at the time their second child, a girl, was born, her state, California, did not yet require routine screening for galactosemia. The baby failed to gain weight and was sent for extensive testing. Galactosemia was finally diagnosed when she was one month old. The disease process was arrested then as all dairy products were removed from her diet, but damage from one month's lost time left her with cirrhosis (hardening) of the liver and cataracts, along with some learning disabilities. At ten she is having difficulty learning to read. Her mother hopes screening will soon be mandatory in all states. She says, "I don't think a parent should have to go through the loss of a child in order to save the next one."

"One third to one half of the families that have a galactosemic child eventually diagnosed have already had one infant die with a disease that, in retrospect, at autopsy or clinically, looked like galactosemia," said Dr. Richard Koch, Professor of Clinical Pediatrics at the U.S.C. School of Medicine and Head of the Division of Medical Genetics at the Children's Hospital of Los Angeles. I asked him how many galactosemic patients were being discovered. "We're running about one in 40,000 here in California," he replied. "It is slightly more common in Dutch and Northern European groups. Galactosemia occurs in blacks at about the average incidence." In discussing the cost, Dr. Koch said, "All you have to do is find one patient, and it pays for all the screening."

The best way to get a law passed requiring that all newborns be tested for galactosemia in your state is to write to your own state representative and state senator asking them to introduce a bill.

In California the incidence of galactosemia has been about one in 40,000. If your state has 400,000 births a year and it doesn't screen for galactosemia, you could be sure of saving ten children from mental retardation and death—if you could get the mandatory testing law put into effect.

You can make a difference.



The blotter paper (right), containing baby Seigel's blood, will be sent to a laboratory to rule out the possibility of PKU and hypothyroidism. The same sample *could* be used to test for a galactose defect. In California, Ohio, Texas and Massachusetts it would be. What about your state?

Attm #3
2-29-83

KANSAS DEPARTMENT OF HEALTH AND ENVIRONMENT

TESTIMONY ON HOUSE BILL NO. 2864

FEBRUARY 29, 1984

HOUSE PUBLIC HEALTH AND WELFARE COMMITTEE

This is the official position taken by the Kansas Department of Health and Environment on House Bill 2864:

House Bill 2864 adds the screening for galactosemia to the newborn screening law first enacted in 1965 to screen for phenylketonuria and amended in 1977 to add congenital hypothyroidism. The law requires the department to conduct educational programs, provide recognized screening, diagnostic and treatment control tests, maintain a registry of cases, and provide the necessary treatment product for diagnosed cases of phenylketonuria, hypothyroidism, and such other diseases as may be appropriately detected with the same procedures for which laboratory services are required.

Infants born with galactosemia have an intolerance to the sugars, lactose and galactose, as the result of one of two enzyme deficiencies, galactose - 1 - phosphate uridyltransferase, or galactokinase. The transferase deficiency is the most common and the most severe, ofter resulting in early death of the infant. Symptoms usually do not appear until the infant starts milk feedings since the principal sugar in milk is lactose. The abnormality is manifest in organ systems by the following signs and symptoms: the eye - lenticular catarracts; the liver - hepatomegaly, jaundice, hypoglycemia; the kidney - generalized aminoaciduria; the gastrointestinal tract - anorexia, vomiting, abdominal distention, ascites, cholelithiasis, acholic stools; the central nervous system - lethargy, hypotonia, mental retardation. Mental retardation appears to be irreversible when it occurs.

Treatment of the disease is restriction of foods containing lactose and galactose from the patient's diet the remainder of life in the majority of cases. With this treatment measure, largely the replacement of milk and milk products with other nonlactose containing nutrients, the child can develop normally and lead a normal life. Otherwise the outcome can be devastating to the child and family.

The disorder is transferred as an autosomal recessive trait. The prevalence of the disorder is 1:50,000.

Over 30 states have added the test for galactosemia to the basic screening tests for phenylketonuria and hypothyroidism and a number of states have pilot programs to evaluate the usefulness of adding this test to the general screening program. In Kansas by adding this screening procedure the Kansas Department of Health and Environment laboratory would require additional staffing and funds for equipment and supplies. Approximately 1/3 of newborns in Kansas have genetic screening performed by eight hospital laboratories and one independent laboratory rather than the State Health Department laboratory. A recent survey of these hospitals indicated that they would cooperate in testing for galactosemia if it became a required procedure for routine screening.

Attm #3
2-29-1984

There would be a problem for the Kansas Department of Health and Environment laboratory as well as private laboratories to get the test started up and on-line before January 1, 1985. It would be preferred that should this test become a requirement for newborns in Kansas that the requirement begin January 1, 1985. There would be initial start up costs for equipment and supplies and an ongoing cost of laboratory personnel and follow-up in educational activities which would continue after the first year.

The Department of Health and Environment recommends that this bill be recommended favorably for passage provided adequate resources are identified to carryout the testing and the follow-up.

Presented by: Virginia E. Tucker, M.D., Medical Director
Crippled and Chronically Ill Children's Program
Kansas Department of Health and Environment

Attm # 4
2-29-84

KSNA

the voice of Nursing in Kansas

Statement of Kansas State Nurses' Association by Evelyn Smith, R.N. before the House Public Health & Welfare Committee
February 29, 1984
Supporting HB 2864 Requiring Diagnostic Test on Infants to Determine If They Have Galactosemia

Mr. Chairman and members of the committee, my name is Evelyn Smith. I am a family nurse practitioner as well as an educator at Wichita State University. I represent the Kansas State Nurses Association. Galactosemia, though rare, is a diagnosable genetic disease in newborns. Because time is of essence and milk creates problems for these babies, screening may be a lifesaving procedure. I have written an article which includes a case study of such an infant. With proper diagnosis and elimination of milk products those infants may live a fairly normal life.

There are 30 states already routinely screening babies for galactosemia as of April 1983. The cost of screening may range considerably depending on whether it is done on a single basis or multiple screening. From the Infant Screening article, you will note that there are government grants available which encourage such screening.

As I stated in 1980 routine screening is necessary to diagnose galactosemia in the newborn infant before the infant becomes ill. From the research I have done I would personally support such screening in the state of Kansas.

Attm. # 4
2-29-1984

GALACTOSEMIA: AN INBORN ERROR OF METABOLISM

Evelyn J. Smith, R.N.,C., B.S.N.

As a clinical entity, galactosemia was first described in 1908. Not until 1956 was it defined as resulting from a specific enzyme defect. We now know enough about it to describe its mechanism of action, as well as how its structure and function are modified by its various structural gene mutations. "Galactosemia is an inborn error of carbohydrate metabolism in which the body is unable to utilize the sugars galactose and lactose."[1] The reason for the inability to utilize the sugars is an absence of the enzyme galactose 1- phosphate uridyl transferase in the liver.

Normally in liver cells appropriate enzymes are available to promote interconversions between the monosaccharides. The final product is glucose, the sugar that can be utilized by the cells of the body for energy. In galactosemia, one step in that conversion process is absent due to a gene mutation which is responsible for the absence of galactose 1- phosphate uridyl transferase (Gal 1-PUT). As a result galactose circulates unusable in the blood and is excreted in the urine. Cataracts, brain damage, and hepatomegaly with fatty metamorphosis in fatal cases occur.

In an autosomal recessive inheritance pattern the parents must be heterozygous for the trait and usually do not display any symptoms; they are carriers. Males and females are affected with equal frequency. One fourth of the children of parents who are unaffected will be affected. A negative antecedent family history is the usual pattern.

Symptomalogy

Infants appear normal at birth but within a few days they begin to vomit and lose weight. Jaundice and liver enlargement are also early signs. The baby soon appears sleepy, exhibiting nausea, vomiting and diarrhea. Septisemia appears in a few days. Clinical symptoms may lead to severe mental retardation and cataracts, as well as cirrhosis of the liver, if the infant survives but remains untreated.

Diagnosis

The primary health care provider must rule out neonatal jaundice, kernicterus and hypoglycemia when confronted with an enlarging liver. Then studies of galactose 1- phosphate uridyl in RBCs and laboratory levels of galactose in the blood stream or urine should be obtained. Levels of galactose in the urine may not be elevated right away so one must not depend on one lab test alone or discontinue pursuit of the problem when dealing with a sick infant.

Case History

A boy was delivered at term, weighing 3.4 kg, after an uneventful pregnancy. He was the second child in the family. The first child had failed to thrive and died at the age of one month from a generalized fulminating septisemia. The second baby was well until day six, when he fed poorly, failed to gain weight and was found to have a *Klebsiella* septisemia. He improved, over the next week on antibiotics. During this time, however, he became jaundiced with a maximum serum bilirubin (5.85 mg/100 ml) and his liver became palpable, 2 cm below the right costal margin. He had recurrent thrush infections and a low serum IgA. At five weeks, he developed diarrhea; stool cultures yielded *Alkalescens dispar* but other cultures were negative and he improved once again on antibiotics. His hepatomegaly was 3 cm below the right costal margin at six weeks, but he was reported to be feeding well on Ostermilk®. A urine report at that time was negative for reducing substances. At eight weeks he was said to be feeding well and gaining weight, but a Benedicts' test for urine reducing substances was again negative. At nine weeks he was well, but hepatomegaly persisted. Urine was tested again, and showed a trace of galactose with amino-aciduria. A red cell assay for Gal 1-PUT, showed no enzyme activity. He was started on a galactose free diet and progressed well. He is now two years old and making normal developmental progress with no evidence of cataracts.[2]

This child has classic galactosemia. The diagnosis was missed until he was twelve weeks old. The limitations of urine screening tests are obvious. A wider use of qualitative enzyme assay needs to be utilized.

Diagnosis

The urine test may continue to be the basic screening test for galactosemia, but is obviously of no value in an infant who is not feeding regularly on milk. In a sick infant a negative result for reducing substances should not be used alone to exclude galactosemia, in the face of clinical symptoms. In all cases of doubt, an enzyme screening test should be performed.

Beutler and Baluda devised a screening method in 1966, using filter paper with blood spots similar to PKU testing. Further screening for Gal 1-PUT requires ultra-violet light to show the presence of the enzyme in red cells. With classic galactosemia the red cells produce no fluoresence.

Routine newborn screening is necessary for the diagnosis of galactosemia. The full impact of galactosemia of the newborn infant has become evident only since routine screening has been initiated in some states. It is now apparent that death associated with bacterial sepsis may occur in about 30 percent of those with untreated classic galactosemia. This usually occured by the second week of

life.[3] Further procedures are: urine should be tested for reducing substances; if urinary reducing substance is found, galactosemia should be presumed. If urinary reducing substance is found, milk feedings should be discontinued and blood and urine specimens sent to the lab for confirmatory testing. If the infant is ill, bacterial cultures should be obtained and treatment for sepsis initiated. If galactosemia is confirmed treatment should be continued.

Treatment and Counseling

When positive results are found, patients and their families need to be investigated further by a quantitative assay for Gal 1-PUT, so that counseling can be initiated. Then, if prenatal diagnosis is considered, a precise knowledge of familial genotypes is available.

Any one of several phenotypes of galactosemia is likely to be found in the infant with an abnormal screening test. Phenotyping is important as the genetic form of galactosemia may determine the prognosis, course of treatment, and need for genetic counseling.

Treatment of the infant includes galactose free formula, called Nutramigen®, or meat-base formula. Nutramigen is a protein hydrolysate process formula produced by Mead-Johnson. The various formula companies usually have their own brand of meat-base formula. The nurse, in counseling parents about their baby's condition, should suggest several, and know the most economical way of purchasing, as this special formula is more costly than regular formula. If the clinician presents an economical route of purchase, compliance may be higher. As baby grows, parents must be educated, not only to avoid giving their child milk, but to carefully omit any foods containing milk. Some foods have dry skim milk added. Among those are most breads, crackers, pastries and cakes. Butter, margarine and cheese should also be avoided. Brain tissue contains galactose and must be omitted. Labeling must be checked and if a product is not clear, it should be avoided. Parents may have to change their food preparation and eating style somewhat to incorporate their child's needs into their own diet. Processed, quick foods may not be feasible for such families. A good nutritious variety of food can be offered with good planning. The nurse can fill this role of educating parents by taking a family nutritional history and giving lists and suggestions of how to adapt to their child's diet. Parents must be cognizant of the consequences if they fail to follow the prescribed diet. They must understand that this child will *never* be able to tolerate milk or milk products. Depending on a diet without milk, calcium and vitamins need to be calculated on the child's needs, especially vitamins A and D.

As soon as the parents have adjusted to the shock and crisis of having a "different" child, they will become concerned about having more children with the same problem. As discussed before, galactosemia is an autosomal recessive disease and parents have a 25 percent chance of having another child with galactosemia and that 50 percent of the children will be carriers. Other alternatives to having their own children should be offered. Prenatal diagnosis is an option for these parents, and should be discussed. Include where it can be done, how much it costs, risks involved, and what is usually expected of them if a positive report is received. Justification of amniocentesis may arise, since these children can be treated. The procedure can remove uncertainty in the minds of parents. Further, knowledge that an affected fetus is to be delivered insures immediate, proper treatment.

Even with prompt diagnosis and treatment of the disease, these children may still be undersized and of low intelligence.[4] They may have coordination and perceptual disabilities which are thought to be associated with their low intelligence. One study indicated such children had more emotional disturbances and were particularly sensitive to criticism. They tended to have poor relationships with the adult world.[4] Parents will continue to feel frustrated by their child's problems. Referral may be needed at some time for family counseling. The nurse can help parents set realistic goals for their child on the basis of individual testing of capabilities, usually done through the public schools. Assessment of vision and fundoscopic examinations to rule out cataracts are essential in follow up care.

As the child enters adolescence and even before, he must also be educated about his or her condition to prevent noncompliance of diet. This is a common problem in pediatrics. Children at this stage do not want to be different. As the child grows into adulthood, further genetic counseling needs to be done regarding marriage and children. (NP)

REFERENCES

1. Whaley, L.: **Understanding Inherited Disorders**, St. Louis, C.V. Mosby, 1974.
2. Monk, A., Mitchell, J., Milligan, W. and Holton, J.: "Diagnosis of Classical Galactosemia," **Archives of Disease in Childhood**, 52, 1977, pp. 943-946.
3. Levy, H. and Hammersen, G.: "Newborn Screening for Galactosemia and Other Galactose Metabolic Defects," **The Journal of Pediatrics**, 92:6, 1978, pp. 871-877.
4. Lee, D.: "Psychological Aspects of Galactosemia," **Journal of Mental Deficient Research**, 16:173, 1972, pp. 173-191.

RELATED READINGS

Beutler, E.: "Screening for Galactosemia," **Israel Journal Medical Science**, 9:9-10, 1973, pp. 1323-1329.

Gershen, J.: "Galactosemia: A Psychosocial Perspective," **Mental Retardation**, Aug. 1975, pp. 20-23.

Guyton, A.: **Textbook of Medical Physiology**, (4th Ed.) Philadelphia, W.B. Saunders, 1971.

Hudson, A.: "Galactosemia: Screening for Inform Errors of Metabolism in Infancy," **Nursing Mirror**, Aug. 28, 1975, 141, pp. 64-66.

Kelly, S., Desjardins, L., Armerding, P. and Burns, J.: "Phenotypes of Galactosemia in Infants Screened at Birth," **Journal of Medical Genetics**, 10:27, 1973.

Mercer, R.: "Crises: A Baby is Born with a Defect," **Nursing '77**, Nov. 1977, 7, pp. 45-47.

North, A.: "Screening in Child Health Care: Where Are We Now and Where Are We Going?" **Pediatrics for the Clinician**, 54:5, Nov. 1974, pp. 631-640.

Perry-Millers: **Pathology, A Dynamic Introduction** (3rd Ed.), Boston, Little, Brown and Company, 1978.

Tedesco, T.: "Studies and Molecular Defect in Galactosemia," **UCLA Forum in Medical Sciences**, 18, 1975, pp. 467-476.

Tedesco, T., Morrow, III, G. and Mellman, W.: "Normal Pregnancy and Childbirth in a Galactosemic Woman," **Journal of Pediatrics**, 81:6, 1972, pp. 1159-1161.

Tedesco, T., Wu, J., Boches, F. and Mellman, W.: "The Genetic Defect in Galactosemia," **Physiology in Medicine**, 292:14, 1975, pp. 737-740.

Won, G., Donnell, G., Bergren, W., Alfi, O. and Golleus, M.: "Prenatal Diagnosis of Galactosemia," **Clinica Chimica Acta**, 74, 1977, pp. 227-235.

To enhance the development of babies...
INFANT SCREENING
Volume 5, No. 1, April, 1983

PROFILES IN PREVENTIVE MEDICINE

Editor's Note

Good medicine cures, great medicine prevents disease. Yet the pioneers who developed preventive medicine in the field of infant screening fought a hard battle against medical forces still wedded to the treatment-for-service and cost-effective approaches in pediatrics. Profiles of these farsighted pioneers will be presented in the newsletter from time to time. We are pleased to initiate the series with a profile of a great innovator, Dr. Robert Guthrie, who launched the program of infant screening in medicine.

Dr. Robert Guthrie

"If I have made any significant contribution to science and human betterment, it was my development of screening tests to detect rare metabolic conditions in infants, such as phenylketonuria (PKU)." (Blatt, in press) Thus, Dr. Robert Guthrie begins a chapter about himself and his PKU test in a book about the lives of people who contributed to the field of mental retardation. His chapter is entitled "Explorations in Prevention".

After growing up in a small town in Marianville, Missouri, Guthrie went to the University of Minnesota and earned a bachelors degree, a Ph.D. and an M.D. He also holds a masters degree from the University of Maine. He never practiced medicine, though, choosing instead to be a medical researcher.

In 1946, Guthrie began working at the National Institutes of Health as the only scientist working there in biochemical genetics, a new field at the time. In 1951, Guthrie moved his family to Staten Island and began working at the Sloan Kettering Institute for Cancer Research in New York City on problems related to childhood leukemia. While living on Staten Island, his six year old son was diagnosed as being mentally retarded.

In 1954, Guthrie moved to Buffalo, New York, to join the staff of the Roswell Park Memorial Institute, to continue his cancer research, specializing in microbial biochemical genetics. Buffalo's Children's Hospital asked Guthrie to develop a method of measuring blood phenylalanine to help with the newly developed treatment for phenylketonuria. He applied similar methods he was already using in biochemical genetics to quickly develop a bacterial test to measure blood phenylalanine quantitatively. This was done by combining bacillus subtilis in a simple agar culture medium with an available chemical antagonist of phenylalanine. By placing paper disks that had been punched from a filter paper impregnated with a measured amount of blood serum, he could estimate the amount of phenylalanine by comparing the diameter of the growth zones. The test was easy to perform since it only required a tiny amount of blood from the child.

Dr. Guthrie then moved over to the Department of Pediatrics at Children's Hospital and began studying the different microbial inhibitors available that had been synthesized for use in cancer chemotherapy. He hoped he could use this approach to find ways of detecting new metabolic diseases in children.

After he started this program in 1958, an amazing coincidence occurred. Guthrie learned that his wife's niece had just been diagnosed as having phenylketonuria. However, because she was not diagnosed until she was 14 months old, she had already become severely retarded.

At this point, Guthrie conceived the concept of applying the tests they already were using to monitor blood phenylalanine specimens from older children to newborn babies. If every baby could be tested, then the children who had phenylketonuria could be detected early and in time for treatment to prevent the otherwise almost inevitable mental deterioration. As Guthrie writes, "as it turned out, the test was even better than I would have dared to dream. The amount of blood on each one fourth inch (7 mm.) disk, punched with a paper punch from drops of blood spotted on filter paper, varies no more than approximately 3 to 5% in dry weight from one disk to another. This meant that an adequately quantitative sample could easily be obtained, similar to the liquid blood measured in a micro pipette in the usual way...Curiously enough, I have discovered as the years have gone by that no one, as far as I have been able to determine, in the entire history of medicine, has used dried spots of blood on filter paper, punched to obtain a quantitative sample of testing for any purpose."

Continued on page 2

Screening Status Report

The status of screening programs across the country is constantly in the process of change. This is a state-by-state report, utilizing the most current information available. (Black dots are added to the list below a specific test only if every baby in a particular state is being screened for that particular disease. We would appreciate information from the individual state informing us when it is necessary to add or subtract a dot.)

	PKU	T-4	MSUD	Homocyst.	Galactosemia	Tyrosinemia	Sickle Cell	Other
Alabama	●	●					*	
Alaska	●	●	●	●	●	●		
Arizona	●	●	●	●	●		●	
Arkansas	●	●					●	
California	●	●			●		*	
Colorado	●	●	●	●	●		●	
Connecticut	●	●	●	●	●	●		
Delaware	●	●	●	●	●			
D.C.	●	●	●	●	●			
Florida	●	●	●		●			
Georgia	●	●	●	●	●	●	*	●[1]
Hawaii								
Idaho	●	●	●	●	●	●		
Illinois	●	●						
Indiana	●	●						
Iowa	●	●	●		●			
Kansas	●	●						
Kentucky	●	●			●			
Louisiana	●	●						
Maine	●	●	●	●	●			
Maryland	●	●	●	●	●			
Massachusetts	●	●	●	●	●			
Michigan	●	●						
Minnesota	●	●			●			
Mississippi								
Missouri	●	●						
Montana	●	●	●	●	●	●		
Nebraska	●							
Nevada	●	●	●	●	●	●		
New Hampshire	●	●	●	●	●			
New Jersey	●	●						
New Mexico	●	●	●	●	●		●	
New York	●	●	●	●	●		●	●[2] ●[3]
North Carolina	●	●						
North Dakota	●	●						
Ohio	●	●		●	●			
Oklahoma	●	●						
Oregon	●	●	●	●	●	●		
Pennsylvania	●	●						
Rhode Island	●	●	●	●	●			
South Carolina	●	●						
South Dakota	●	●						
Tennessee	●	●						
Texas	●	●		●	●			
Utah	●	●			●			
Vermont	●	●			●			
Virginia	●	●						
Washington	●	●						
West Virginia	●	●			●			●[1] ●[2]
Wisconsin	●	●	●		●			
Wyoming	●	●	●	●	●		●	

*(on request)
[1]Hemoglobinopathies
[2]Adenosine-deaminase
[3]Histidinemia
(Alabama, Tennessee / at conference update)

Continued from page 1

The rest is history. The first pilot study began the summer of 1961 and, as a result of the screening, 23 cases of phenylketonuria were discovered at Newark state school. Actually, the first non-selective infant screening was started in a small hospital in Jamestown, New York. Then with funds from the Children's Bureau, Dr. Guthrie and his team rented a house close to Children's Hospital and converted it into a miniature factory to prepare enough test kits so that screening could start in 29 states. They prepared materials for a million tests. They set up quality control procedures to insure the materials were uniform. In the first countrywide testing of 400,000 babies who were screened, 39 cases diagnosed as PKU were detected.

Guthrie reports "Many of the doctors in the United States were not interested at all in the test. In fact, they were often very antagonistic to laws requiring the test. In all states except Massachusetts, state medical societies resisted the idea of a law, if they acted on the issue at all. Much of this attitude is related to the way in which our health care is organized. Much of the controversy concerning PKU screening and treatment was stimulated by resentment against the laws that were passed. Laws requiring babies to be tested were considered by many doctors as interference, an invasion by the government in a private medical matter. The rights of doctors seemed to be more important to some than the rights of infants to be protected from mental retardation."

Today, the battle for phenylketonuria appears to have been won in the United States (see Screening Status Report, on left) and also in many countries around the world. In the years since PKU testing was developed, 30 new tests have been devised in Guthrie's laboratory. Many of these tests are now used in newborn screening programs in areas of the United States and the world.

But Guthrie is not resting on his laurels. He is currently working towards a law to prevent children from becoming mentally retarded because of lead poisoning. In addition to his other duties for the past 12 years, he has been an integral part of a national team, pushing people to realize that lead poisoning is a major health problem in the United States. In 1980, he began testing lead specimens from children in the New York State area and found four children with lead levels so high they had to be hospitalized.

Thus, this indefatigable man is now lobbying against an environmental hazard, continuing his commitment to the most helpless members of our society—infants and young children.

Throughout the years since Dr. Robert Guthrie began promoting screening of infants for metabolic errors, he and his associates at the State University of New York at Buffalo have been most helpful to screening programs by providing assistance with quality control and procedure development. A grant from the U.S. Maternal and Child Health Service was renewed specifically to encourage and assist screening laboratories to expand their programs beyond PKU and hypothyroidism to include galactosemia, maple syrup urine disease, homocystinuria, and sickle cell disease.

For further information call 716-831-2351 or write to Dr. Guthrie at Acheson Hall, Room 352, 3435 Main Street, Buffalo, N.Y. 14214. Drs. Guthrie, Edwin Naylor, David Jinks and Mrs. Sally Bloom are willing to help with any problems.

Attm. #4
2-29-84

KANSAS
ASSOCIATION FOR RETARDED CITIZENS, INC.

11111 WEST 59th TERRACE
SHAWNEE, KANSAS 66203
(913) 268-8200

To: House Public Health and Welfare Committee — 2/29/84
Marvin Littlejohn, Chairman

From: Kansas Assn. for Retarded Citizens — Re: HB 2864
Lila Paslay, Volunteer

It is a special privilege to come before you in support of a proposal through which one of the causes of mental retardation may be prevented. We in the Kansas ARC have long been proud to be from a state which has been among the leaders in applying technologies toward prevention which have evolved through research. Kansas was among the first to require testing and diet for children with PKU, among the first to require immunization against red measles and German measles.

We agree with a former Assistant Surgeon General, Dr. Albert Chapman who said "millions are spent in this country annually for medical research, but only pennies are spent for applying the findings....Spending millions for research and pennies for application is like buying bread and not eating it."

In the Report of Task Force on Law and Mental Retardation, Judge David L. Bazelon, Chairman, 1963: "The long-run problem for justice in regard to retardation is to ensure that every American child has the opportunity to <u>be</u> "created equal" in the sense that he be neither born so badly that his equality is destroyed before he comes into it, nor born into such circumstances that the promise of his equal birth is broken before his life is fairly begun. The state cannot assure a child a good set of genes.....But to fail to supply, as quickly as possible, as specifically as possible and as effeciently as possible

Attm. #5
2-29-1984

any reasonable medical, social or legal remedy for retardation is to impose upon a child the greatest injustice of all."

We believe the addition of galactosemia to the act for screening, diagnosis, and treatment control tests is indeed a reasonable medical remedy for a condition that cuases mental retardation and we urge your support of such legislation.

We as parents are extremely grateful to the many physicians, teachers, and other professionals who labor for each small gain with our retarded sons and daughters of today. But we cannot help but think now and then of what might have been, as well as hope of what may yet be. And so we work for progress in behalf of those whom we know and love so well.

But we also work in behalf of those, who, if we are successful in both research and prevention we will never need to know as mentally retarded.

We support HB 2864 as one more step in preventing one more cause of mental retardation, and urge your favorable response to it.

Attm 6
2-29-84

Kansas Advocacy & Protective Services for the Developmentally Disabled, Inc.

kaps

Suite 2, the Denholm Bldg.
513 Leavenworth
Manhattan, KS 66502
(913) 776-1541

To: The House Committee on Public Health and Welfare
Representative Marvin L. Littlejohn, Chairperson

From: Kansas Advocacy and Protective Services
R.C. Loux, Chairperson

Date: February 29, 1984

Re: H.B. 2864

Galactosemia is a metabolic inability to convert galactose to glucose. Left untreated, infants may die. Survivors of this condition can be mentally retarded and have other serious impairments.

Early diagnosis and treatment is imperative. The prognosis is good if galactose is eliminated before birth and until age 6.

Passage of H.B. 2864 will make it possible for Kansas to make wise use of the medical knowledge we now have available about galactosemia. Through effective use of education, screening, diagnosis and treatment we can prevent serious handicapping conditions from occuring in the future.

Respectfully submitted,

Joan Strickler
Executive Director

JS/jw

Attm. #6
2-29-1984

Attm. #7
2-29-84

KSNA

the voice of Nursing in Kansas

Statement of Kansas State Nurses' Association
by Diane Bottorff, R.N., Assistant Director
before the House Public Health & Welfare Committee

February 29, 1984

In strong support of HB 3024 Prohibiting Home Health Aides from Prefilling Insulin Syringes

Mr. Chairman and members of the committee, I am Diane Bottorff, an Assistant Director of the Kansas State Nurses' Association. KSNA speaks in strong support of HB 3024. Having over six years of experience in home health nursing, part of that time as a supervisor of home health aides, I feel well qualified to address the content of this bill.

KSNA has long held the position that medication administration is the function of licensed nursing personnel. In relating that position to the content of this bill, we believe that prohibiting home health aides from prefilling insulin syringes is necessary for several reasons.

First, home health aides are not required by all home health agencies to have completed a nurse aide course. Thus, some have had no formal training in the care of the sick and disabled. Even those who have taken a formal course have no training related to drugs. Home health agencies are not permitted by Medicare regulations to allow aides to administer medications in clients in their homes. Permitting home health aides to prefill insulin syringes is at best risky due to their lack of preparation for such a responsibility.

A second point is that while the Nurse Practice Act allows for delegation of tasks by licensed nurses to auxiliary nursing personnel under their supervision, in the home setting the aide and the nurse are seldom present at the same time. No one would be observing to make sure that she was accurately filling the insulin syringes. Yet the licensed nurse would be liable for any error which might occur.

Finally, insulin is a potentially dangerous drug. Putting too much or too little insulin in the syringe can lead to severe complications or could even be fatal. In addition, management of

of a diabetic client entails more than just filling syringes. The licensed nurse also assesses the diabetic's overall status as it relates to diet, exercise, the response to insulin and other drugs being administered, and the effects other disease states may have on the diabetes. The home health aide would not have the knowledge base to do an assessment of this type and certainly would not be expected to have the judgment to determine when the insulin dose is no longer appropriate. The diabetic who needs insulin syringes prefilled is most vulnerable due to failing eyesight, poor coordination or both. This person may be living alone and dependent on outside sources to assess his condition and detect possible complications.

In summary, KSNA speaks in support of HB 3024 because it prohibits unlicensed, minimally trained persons from performing a task for which they are not prepared. It keeps issues dealing with medications within the scope of practice of licensed persons and ensures that clients particularly the most vulnerable will be managed by those who are best qualified to do this.

Attm #7
2-29-1984

Attm. #8
2-29-84

THE KANSAS PHARMACISTS ASSOCIATION
1308 WEST 10TH
PHONE (913) 232-0439
TOPEKA, KANSAS 66604

KENNETH W. SCHAFERMEYER, M.S., CAE
PHARMACIST
EXECUTIVE DIRECTOR

STATEMENT TO THE HOUSE COMMITTEE ON PUBLIC HEALTH AND WELFARE

SUBJECT: House Bill 3024 - Prefilling of Insulin Syringes by Home Health Aides

Mr. Chairman and Members of the Committee:

I am Ken Schafermeyer, Executive Director of the Kansas Pharmacists Association, an organization representing approximately 80% of the practicing pharmacists in the state of Kansas. Thank you very much for the opportunity to address you on HB 3024 which would prevent home health aides from prefilling insulin syringes.

We urge your support of this bill. Although insulin may be purchased without a prescription, it can be a very dangerous drug if used improperly. There are 31 varieties of insulin (e.g. rapid acting, intermediate acting, and long acting). Even the most expert clinician has to work hard to keep current on these insulins. Often a patient may be required to mix two different types of insulin in the same syringe. A patient may also have two insulin injections daily using one type of insulin in the morning and a different type, or quantity, in the evening.

The amount of insulin, the type of insulin and the timing of the injection is extremely important and must be customized for each patient. Too much insulin can result in a severe reaction known as "hypoglycemia," or "insulin shock." Insulin shock is a serious emergency and may result

attm. #8
2-29-1984

in brain damage, stroke and even death if untreated. Too little insulin can result in "hyperglycemia" which can also be dangerous.

Insulin is classified as an over-the-counter (OTC) drug, not because it is not dangerous, but because it must be readily available to diabetics who need it. Because insulin is not a prescription drug, current law does not prohibit under-qualified persons from prefilling these syringes. It is too easy to contaminate the sterile solution, the syringe or the needle. It is too easy to fill a syringe with the wrong type or amount of insulin. Diabetics are trained to administer injections to themselves but home health aides, who may know nothing about diabetes, insulin or patient care, cannot be expected to keep each patient's individual medication needs in mind. He or she cannot be expected to realize the inherent dangers in preparing these medications.

Therefore, Mr. Chairman and members of the Committee, I urge your support of HB 3024 which would prohibit unlicensed persons from prefilling insulin syringes. Thank you.

Attm. #9.
2-29-84

K.S.A. 65-509

Section 1. (a)Except as otherwise provided in this section:

(1) No person shall advertise that such person will adopt, find a home for or otherwise place a child;

(2) No person shall offer to adopt, find a home for or otherwise place a child as an inducement to a woman to come to such person's maternity hospital or home during pregnancy or after delivery; and

(3) No person shall offer to adopt, find a home for or otherwise place a child as an inducement to any parent, guardian or custodian of a child to place such child in such person's home, institution or establishment.

(b) The provisions of paragraph (1) of subsection (a) of this section shall not apply to a licensed child placement agency operating as authorized by KANSAS law.

(c) As used in this section:

(1) "Advertise" means to communicate by newspaper, radio, television, handbills, placards or other print, broadcast or electronic medium;

(2) "person" means an individual, firm, partnership corporation, joint venture or other association or entity.

Attm. #9
2-29-1984