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ABSTRACT

This booklet summarizes current knowledge about phenylketonuria (PKU), an inherited condition that results in severe mental retardation if untreated, and discusses the psychoeducational implications of the condition. The introduction stresses the importance of early diagnosis (during the first days of life) and the effectiveness of a diet that avoids the amino acid phenylalanine in preventing mental retardation. Individual sections summarize information about screening, diagnosis, and incidence; diet management; the role of school personnel; the elementary student with PKU; and the adolescent student with PKU. The booklet outlines four major responsibilities of teachers, counselors, administrators, and staff when a PKU child enrolls in their school: (1) treatment of the child as a normal, healthy student; (2) establishment and maintenance of open communication with the student and parents in order to facilitate good dietary management; (3) encouraging the adolescent student to continue the restricted diet; and (4) identification of students with possible PKU who are not on the special diet. A glossary defines key terms. A list of resources includes clinics for treatment, materials for parents of young children with PKU, booklets and videos explaining PKU to children and youth, materials concerning maternal PKU, references for teachers and other professionals, and references in the medical literature. (Contains 25 references.) (DB)

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Education of Students with Phenylketonuria (PKU)

**Information for teachers, administrators
and other school personnel**



U.S. DEPARTMENT OF HEALTH AND HUMAN SERVICES
Public Health Service
National Institutes of Health

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Special Terms

Three abbreviations are used which may be unfamiliar to the reader:

PKU: the short version for phenylketonuria.

phe: which stands for phenylalanine--the amino acid found in all protein, which individuals with PKU cannot metabolize.

Maternal PKU: the problems associated with pregnancy where the mother-to-be has PKU.

A short Glossary of terms associated with this condition will be found at the end of the booklet.

Preface

The purpose of this booklet is to acquaint teachers, school nurses, administrators and other school personnel with current knowledge about phenylketonuria (PKU), and to identify the psycho-educational implications of the condition. Suggestions are provided to enable schools to educate children with PKU in accordance with the principles embodied in statute and constitutional law which provide for a free, appropriate public education in the least restrictive environment for all children. Resources for further information about PKU and its associated problem, Maternal PKU, are provided at the end of the booklet.

This is the third edition of this publication (previous versions having been published by the U.S. Public Health Service). Like its predecessors, it is based primarily on the knowledge gained from the Collaborative Study of Children Treated for Phenylketonuria, a project started in 1965 as a joint research effort of 15 PKU clinical programs throughout the United States. Coordination of all data collection was undertaken by Childrens Hospital of Los Angeles, and was initially supported by the Bureau of Community Health Services through funds appropriated under title V of the Social Security Act, Grant No. MCT-0000466. The last three years of the project was supported by the National Institute of Child Health and Human Development Grant Number R01-HD0-8543. While the project has officially terminated, data continue to be gathered and analyzed as the study population moves through the school years and into young adulthood.

Most clinics originally involved in the dietary treatment study are now participating in the study of the special problems related to Maternal PKU. This project is also being coordinated through the Childrens Hospital of Los Angeles under contract N01-HD-4-3807, funded by the National Institute of Child Health and Human Development. Five regional contributing centers have been designated to coordinate efforts: four in the United States, and one in Canada. The funding agency for the Canadian project is the National Health Research Development Program. The address and phone number of each of these centers are provided in the "Resources" section at the end of the booklet.

Education of Students with Phenylketonuria (PKU)

Introduction

Phenylketonuria, called PKU for short, is an inherited condition which prevents the affected individual from normally metabolizing--or using--phenylalanine (phe), one of the essential amino acids found in all protein foods. Unless the condition is detected and treatment is initiated soon after birth, this hereditary biochemical abnormality prevents normal brain development and usually results in severe mental retardation. Other manifestations such as skin rash, seizures, excessive restlessness, irritable behavior and a musty body odor may also be present.

Fortunately, detection of the condition shortly after birth through the use of a routine blood screening procedure has become standard practice in every state and Canadian province since 1991. Placing the baby on a phenylalanine-restricted diet within the first weeks of life, and maintaining good diet control thereafter, is effective in preventing the damaging effects of PKU. Treatment requires elimination of foods naturally high in protein. Special care must be taken to maintain enough--but not too much--phe in the child's diet. Such a balance is obtained only through careful nutritional, biochemical and medical supervision, and considerable effort by the parents.

Beginning at an early age, most children with PKU learn to discriminate between foods that are allowed on their diet from those that are not. However, maintaining the diet may become more difficult when a child with PKU enrolls in school. "Swapping" lunch items may be a temptation; or students may try a diet soft drink which uses the artificial sweetener, NutraSweet (a product containing phe).

While education about the diet is an ongoing process in the homes of most children with PKU, educators need to be familiar with PKU in order to be supportive to the child and the parents, and to assist with diet management during school hours.

Benefits of Treatment

Untreated or late-treated persons with PKU usually develop mild to severe mental retardation. Therefore, it is imperative that the diagnosis of PKU be established within the first days of life, before the onset of mental damage. All states and Canadian provinces have blood screening programs to detect PKU shortly after birth, and similar programs are in place in many other countries.

The Collaborative Study of Children Treated for Phenylketonuria has demonstrated the value of the phe-restricted diet in preventing mental retardation without affecting physical growth and development (Williamson et al., 1981). When treatment is instituted within the first few days of life, and the diet is maintained as prescribed by a PKU treatment center, normal development can be anticipated (Holtzman et al., 1986).

Like other children, those with PKU contract the usual childhood illnesses, and during periods of high fever, the child's blood phe levels may rise dramatically. It is the general consensus that these deviations from optimal phe levels are best ignored; they are transient, have not proven detrimental, and should not be the source of parental anxiety. Of greater significance are the long-term effects of persistently high blood phe levels due to chronic poor dietary control.

At this time, early treated children with PKU are enrolled in public schools, and many cannot be distinguished from non-PKU children, except by their adherence to a special diet (Azen et al., 1991). However, a number of early and well treated children with PKU have some discrete mild to moderate learning disabilities despite normal IQs. These problems are often in the mathematics area and may be complicated by distractibility and short attention span. The exact number of these learning disabled children with PKU is not yet clear. It is important that these difficulties be identified early so that remedial steps can be taken and

the inevitable frustration and subsequent behavior problems be avoided or minimized. Some of the oldest individuals identified by newborn screening, and compliant to long-term phenylalanine restrictions, have graduated from high school and are successfully pursuing college educations (Koch et al., 1985; Fishler, et al., 1987; Fishler et al., 1989).

During the 1960s and 70s, it was common practice to discontinue diet therapy during childhood. In the 1980s, data suggested that there were IQ changes in children in the Collaborative Study of Children Treated for Phenylketonuria who had discontinued the phe-restricted diet (Koch et al., 1987). Therefore, it is now believed that the continued use of the phe-restricted diet is indicated throughout school and indefinitely. It has been established that women with PKU who are not on the diet will have babies with birth defects and mental retardation (Lenke and Levy, 1980). The Maternal PKU Collaborative Study is locating young women with PKU reaching child-bearing age to inform them of this problem and to encourage them to stay on (or to return to) a restricted-phe diet, and thus reduce the possibility of damage to their unborn child (American Academy of Pediatrics, 1985; Koch et al., 1991).

Screening, Diagnosis and Incidence

After its discovery in 1934 by Dr. A. Folling in Norway, and until the early 1960s, PKU was detected with a urine "wet diaper" test. This method has many disadvantages, and since 1964 has been replaced by a blood test which can be administered with great accuracy as early as the first few days of life. Infants initially screened before 24 hours of age should be rescreened by three weeks of age (American Academy of Pediatrics, 1982). When the baby has higher than normal levels of phe in the blood, confirmatory tests must be performed, and if the diagnosis is established, diet treatment begins as soon as possible. Results from the Collaborative Study of Children Treated for Phenylketonuria clearly indicate a loss in IQ scores if the baby is not put on the diet within the first 20 days of life and kept in good dietary control

Approximately five children with PKU are born in the United States each week. This means an estimated incidence of one baby with PKU born in about 15,000 live births.

Diet Management

Proper diet management is essential in the treatment of PKU. This involves severely limiting the child's intake of all foods containing protein. Protein contains the amino acid, phenylalanine (phe), which the child cannot effectively utilize. Milk and other dairy products, meat, fish, eggs, dried beans and peas, and nuts are concentrated sources of phe; fruits, vegetables, and cereals contain smaller amounts. The only foods that do not contain phe are sugar, oil, pure starch and water; and products made with these ingredients such as hard candy and soft drinks. Special medical foods which provide protein and important nutrients are an essential part of the food intake pattern of individuals with PKU. Special low protein foods and baking ingredients are available from several companies (see Resources section at end of booklet).

Normally, the body uses a small part of the phe from dietary protein for growth and repair of body tissue, and changes part of the phe into other useful chemical compounds such as tyrosine. Tyrosine is another amino acid and is needed to make proteins, hormones and neurotransmitters which control brain functions. Tyrosine also helps to make pigments for skin and hair coloring. The enzyme necessary to convert excess phe into tyrosine does not work effectively in the person with PKU. The absence of tyrosine combined with the excess phe and abnormal compounds circulating in the body leads to brain damage. This is particularly true in the first few years of life when dietary compliance is critical to prevent damage and to insure normal growth and development.

Even after reaching school age, continued use of a restricted-phe diet is indicated. The Collaborative Study of Children Treated for Phenylketonuria found that children with PKU who were taken off diet began to show a

drop in I.Q. scores and poorer school performance (Williamson et al., 1981).

The special PKU diet is designed to give each individual the exact amount of phe required for adequate growth and to prevent buildup of harmful amounts. This delicate balance, unique to each person, is achieved by careful measuring of all food and calculating the phe intake on a daily basis. Nutritionists and parents have developed cookbooks and food lists to assist in this exacting task (see Resources section at end of booklet). Frequent blood tests are necessary to monitor the blood phe levels and to determine dietary effectiveness.

In order to supply the other essential elements of protein needed for normal growth and development, a special protein source is necessary. These sources of protein, called medical foods, contain all the amino acids except that most or all of the phe has been removed (see Medical Foods for the names and manufacturers of such products, listed in the Glossary at the end of the booklet). Some of the products have added fat and carbohydrates, while all have been fortified with vitamins and minerals. These medical foods are generally taken as a liquid beverage. The medical food plus carefully selected foods low in protein provide a balanced diet for the child. Often the student with PKU will take the product before or after school, rather than bring it to school.

Special diet management problems may occur at school. For example, young children with PKU cannot have a high protein snack. Special events, such as a birthday, may involve a cake containing forbidden high-phe food ingredients. Because the child with PKU should not eat these foods, he/she may be identified as "different." Parents commonly provide alternatives for their child. However, teachers and administrators must be aware of potential problems and help the child with PKU to participate maximally in all activities, including snack-time and parties by notifying parents to send substitutes.

If a school lunch program is provided, some foods can be safely eaten by the child with PKU. It is often helpful to send the week's

menu home so the parents can indicate which items (and in what quantity) are allowed, and which must be avoided, and return the menu to the teacher. By the time children with PKU reach middle school, and often earlier, they themselves will be able to identify foods that are on their diet. However, they may still require help in determining the appropriate quantity. The ability of children with PKU to participate in the same lunchroom program as their classmates, if appropriate, will lessen the feelings of being different, and thus contribute to the student's healthy social and emotional growth.

Role of School Personnel

As children treated for PKU reach school age, like all children, they move out from the protective environment of their home to a more complex school situation. The child with PKU may wish to experiment with foods that other children are eating. The teacher and other school personnel need to be aware of the potential danger that such innocent food-swapping holds for the child with PKU. While an occasional dietary indiscretion will not cause serious harm, frequent variations--especially when not reported to the parents--could lead to problems which will affect school performance and behavior. An important role for school personnel, therefore, is to work effectively with the parents in maintaining good dietary control.

Children who have been diagnosed early and maintained on an appropriate PKU diet are subject to the same problems, likes and dislikes as other school children of the same age. Variations in height, weight or other physical aspects are probably unrelated to the existence of PKU, or to dietary treatment. The Collaborative Study of Children Treated for Phenylketonuria found that children who have been identified early and satisfactorily treated for PKU have normal growth and development. Likewise, the average IQ scores of children in the study were near the mean score of 100 (Koch et al., 1984). An evaluation of psycho-educational measures, such as the Illinois Test of Psycholinguistic Abilities, the Frostig Visual Perception Test, and the Bender-Gestalt

Test, revealed no special profiles of strengths or weaknesses in children early-treated for PKU. Average school achievement was also near expected levels (Fishler et al., 1989).

It may be desirable for the teacher or other school personnel to provide a direct educational program to explain the child's need for a special diet to other children in the class, and perhaps to the parents of the non-PKU children. Young children can be thoughtless in dealing with a classmate whom they discover is "different" in some way. The teacher's simple explanation of the special diet should help the other children accept it.

As the child grows older, peer pressure to conform with dress standards and social behavior, including eating habits, increases the likelihood that the child will experiment with various foods that may contain excessive amounts of phe. For the adolescent female, remaining on the diet is especially important as she approaches childbearing age. Without restriction in phe-intake before and during pregnancy, a woman with PKU risks extensive damage to her unborn child. Directors of PKU clinics are recommending that females with PKU remain on a low phe diet throughout their school and childbearing years, as it is much more difficult to return to the diet after an unrestricted period (Holtzman et al., 1986). This represents a relatively recent change in policy. Previously, it was thought that the diet could be discontinued soon after the child entered school. Secondary school counselors and guidance personnel can be helpful in reinforcing the need for continued dietary restrictions during the adolescent years and into adulthood.

The Elementary School Student with PKU

By the time the child with PKU reaches school age, parents will have had several years' experience with dietary treatment. The child's medical experiences will be different from his or her peers due to the special diet, periodic visits to the PKU clinic, and frequent blood tests. As a result, it is important for the school to be sensitive not only to the child's diet but

also to emotional and social needs. The child's emotional needs are best met by treating him or her as much like a non-PKU child as possible. However, children with special needs often require a little extra support from parents and teachers to feel accepted and "normal." Based on experience with children in the Collaborative Study of Children Treated for Phenylketonuria, teachers, counselors and administrators in preschool or elementary schools should take special note of the following:

- While children are curious about each other and are quick to note differences such as a special diet, they are also able to understand the concept of a "food allergy." Thus, no attempt should be made to hide the fact that the child with PKU has a different kind of lunch, nor should any lengthy explanation be required at this age.
- Since many schools utilize a "lunch supervisor," that individual should be acquainted with the basic dietary problems associated with PKU. The supervisor should also be aware of the need to avoid excessive attention to the child. Occasional indiscretions are not fatal, and as children with PKU grow older they must take more and more responsibility for their own dietary management. The teacher can assist the child in building the inner controls.
- Some school personnel have been known to overreact to a diagnosis of PKU. One school classified a child with PKU as "orthopedically handicapped" and imposed a limited, adaptive physical education program that was totally unwarranted. Unless the treatment has been initiated late, or the diet not maintained satisfactorily, the child with PKU is not different from other children in physical ability or general health. Similarly, regular school procedures should be followed when a child with PKU becomes ill at school. PKU is not associated with any sudden, dramatic episodes such as the insulin reaction experienced by diabetics, or the grand mal seizure of a child with epilepsy.
- The child with PKU who was diagnosed early and maintained on the special diet will

most often have educational needs that are unaffected by PKU. The child with PKU needs to be treated as any normal child, with strengths and weaknesses, likes and dislikes, and a learning style unrelated to the PKU condition. Most early-diagnosed children with PKU whose diet has been well managed should be able to participate in the regular school program on an age-appropriate basis.

The Adolescent Student with PKU

By secondary school age, the adolescent with PKU should have assumed responsibility for much of his or her own dietary control. For example, he or she might consume some of the PKU medical food products before and after school, and select foods from the school luncheon menu, including salads, vegetables, fruits, and fruit juices (Rees and Trahms, 1986). As indicated earlier, school counselors and teachers should be aware of the recommendation by the Maternal PKU Collaborative Study that a restricted-phe diet be continued indefinitely. It is imperative that women maintain the diet throughout the entire childbearing years (Rohr, 1987).

At the junior or senior high school level, when the curriculum calls for nutrition or cooking classes, it is especially important for the teacher to be in close contact with the parents and the student. They will have, or know of, materials similar to that contained in the Resources section at the end of this booklet. These can be useful supplements to the regular school texts, thus enabling the student with PKU to be included. The need for students with PKU to monitor their diet can serve as a model, especially for children with other kinds of diet problems. Special low protein foods may be baked and served. Science fair projects can be designed by the students to illustrate the genetic and biochemical reactions involved in PKU (see references for teachers in Resources section).

Teachers and counselors in secondary schools can assist the Maternal PKU Collaborative Study in locating young women who were on the PKU diet as a child. This could be done by having the teacher, nurse, physical education

or health instructor note on the health record which students they remember being on the "PKU diet" or a special diet in childhood. If appropriate, the young woman and/or her parents should be urged to call a PKU treatment center in order to obtain important information concerning the effect of PKU on pregnancy (Rohr, 1987; Acosta, 1991).

The Maternal PKU Collaborative Study is designed to locate young women with hyperphenylalaninemia (PKU), to inform them of the problems their babies will have if the PKU is not controlled before and during pregnancy, and to assist them in returning to a phe-restricted diet. Women wishing to return to the diet prior to considering pregnancy, or who are already pregnant, will be enrolled in the study. The pregnancy will be monitored to maintain the safe levels of phe and the other essential elements needed to have a much better chance of having healthy babies. The location of the nearest Maternal PKU Center can be obtained by contacting the appropriate regional Contributing Center of the Maternal PKU Collaborative Study (see list included in Resources section).

Summary and Conclusion

Teachers, counselors, administrators and other school personnel have four major responsibilities when a student with PKU enrolls in their school:

- (1) To treat the student with PKU as a normal, healthy member of the school who is no different from anyone else in physical growth and development, or academic potential.
- (2) To establish and maintain open communication with the student and his or her parents in order to facilitate good dietary management.
- (3) To encourage the adolescent student with PKU to continue the restricted-phe diet and to have it monitored by a PKU center.
- (4) To locate students with PKU who are not on the special diet and encourage them to contact a PKU center.

Glossary

- Amino Acids Organic compounds which combine to form proteins. An "essential" amino acid must be supplied by food; a "non-essential" amino acid can be produced within the body.
- Medical Foods These are special formulas designed to provide the person with PKU with the needed protein, but with little or no phenylalanine. Some of these products and their manufacturers are:
- "Lofenalac" (Mead Johnson), "Analog XP" (Ross Laboratories), and "PKU 1" (Mead Johnson) are commonly used with infants with PKU;
- "Maxamaid XP" (Ross Laboratories), Phenyl-Free" (Mead Johnson), "PKU 2", "PKU 3" (Mead Johnson), and "PKU Aid" (Anglo-Dietetics Ltd.) provide school-aged children with the needed protein source.
- "Maxamum XP" (Ross Laboratories) and "PKU 3" (Mead Johnson) are utilized for pregnant PKU women.
- Enzyme A chemical compound which changes one substance into another (i.e., a catalyst). The enzyme, phenylalanine hydroxylase, is the one that is defective in individuals with PKU--thus they are unable to convert the amino acid, phenylalanine, into other products.
- "Equal" An artificial sweetener containing aspartame, which is 56% phenylalanine (phe) and thus must be totally avoided by persons with PKU.
- Hyperphenylalaninemia The term used to designate a number of conditions (one of which is classical PKU) in which the individual exhibits elevated levels of phenylalanine in the blood.
- Maternal PKU the problems associated with pregnancy where the mother-to-be has PKU.
- "Nutra-Sweet" A sugar substitute found in many "diet" products such as soft drinks. It is made of aspartame which contains 56% phenylalanine and therefore, like "Equal" should not be used by persons with PKU.
- Phenylketonuria An inherited error of metabolism in which the individual cannot metabolize (or use) the essential amino acid, phenylalanine.
- PKU A common abbreviation for phenylketonuria.
- Phenylalanine The essential amino acid in all protein which a person with PKU cannot convert into useful products.
- phe A common abbreviation for phenylalanine.
- Protein Compounds made of amino acids that are essential for all living cells in the body.
- Tyrosine A non-essential amino acid present in all protein foods. In persons with PKU it becomes an essential amino acid because the enzyme necessary for its production from phenylalanine is defective.

Resources

For additional information or assistance beyond that presented in this booklet, the following sources are suggested:

I. Clinics for Treatment of PKU and Maternal PKU:

1. A directory of the 115 PKU clinics in the United States is available in the publication, Schuett, V.E. (1990) National Survey of Treatment Programs for PKU and Selected Other Inherited Metabolic Diseases (DHHS Pub. No. HRS-M-CH-89-5) which can be obtained from:
National Center for Education in Maternal and Child Health
38th and R Streets, N . W.
Washington, D. C. 20057
2. The following regional contributing centers of the Maternal PKU Collaborative Study can also provide information and assistance concerning PKU and maternal PKU:

Northeast Region:

Connecticut, Delaware, New Hampshire, Maine, Maryland, Massachusetts, New Jersey, New York, Pennsylvania, Rhode Island, Vermont, Virginia, West Virginia, District of Columbia.

Director: Harvey Levy, M.D.
Children's Hospital Medical Center
300 Longwood Avenue
Boston, Massachusetts 02115
Phone: 617: 735-6346

Midwest Region:

Illinois, Indiana, Iowa, Kansas, Kentucky, Michigan, Minnesota, Missouri, Nebraska, North Dakota, Ohio, Oklahoma, South Dakota, Wisconsin.

Co-Directors: Reuben Matalon, M.D., Ph.D. and Harvey Levy, M.D.
University of Illinois at Chicago
Department of Nutrition and Medical Dietetics
Mail Code 517, College of Associate Health Professions
Chicago, Illinois 60612
Phone: 312: 996-0995

Southeast Region:

Alabama, Arkansas, Florida, Georgia, Louisiana, Mississippi, North Carolina, South Carolina, Tennessee, Texas, Puerto Rico.

Director: Bobbye Rouse, M. D .
University of Texas Medical Branch
Department of Pediatrics
Galveston, Texas 77550
Phone: 409: 772-2356

Western Region:

Alaska, Arizona, California, Colorado, Hawaii, Idaho, Montana, Nevada, New Mexico, Oregon, Utah, Washington, Wyoming.

Director: Richard Koch, M.D.; Co-Director: Julian Williams, M.D., Ph.D.
PKU Section, Children's Hospital of Los Angeles
4650 Sunset Boulevard
Los Angeles, California 90027
Phone: 213: 669-2152

All Provinces of Canada:

Director: William B. Hanley, M.D.
The Hospital for Sick Children
555 University Avenue
Toronto, Ontario M5G 1X8, Canada
Phone: 416: 598-6356 or 416: 781-1805

II. Materials for parents of young children with PKU:

Parents' Guide to the Child with PKU. (1982, 66 pages: \$8.00)

Topics include understanding PKU, diet management, growth and development patterns, role of others in diet management, management of feeding and behavior problems, management of diet in special situations, future considerations.

Ruth E. Pestle, Ph.D. Florida State University,
Center for Family Study,
103 Sandels Bldg., Tallahassee, FL 32306-2033

Understanding PKU. (1982, 12 pages: single copy free)

Booklet created as a source of basic information concerning PKU.
PKU Clinic, Children's Hospital Medical Center
300 Longwood Avenue, Boston, MA 02115

Living with PKU (1990, 19 pages: single copy free)

A booklet written by parents and staff, including discussion of what PKU is, the genetics and management; interviews with families; and helpful hints from experienced parents.

Mead Johnson
Phone 800: 222-9123

A Babysitter's Guide to PKU. (1986, 8 pages: \$2.50)

An easy to understand pamphlet that helps parents discuss diet management of the child with the babysitter.

Cristine M. Trahms, M.S., R.D.
PKU Clinic, CDMRC WJ-10
University of Washington, Seattle, WA 98195

Babysitters: My Child has PKU. (n.d., 9 pages: single copy free)

Instructions for babysitter regarding diet. Emphasis placed on the fact that the child is like other children except for diet. Brief outline of diet with list of "free" foods.

Karen M. Kinkus, R.D.

Department of Pediatrics, Univ. of Louisville
Louisville, KY 40292

The Child with PKU. (1986, 45 pages: \$5.00)

A booklet for parents; explains PKU, lists sources for information, supplies and assistance.

PKU Section C 19

Child Development, Dept. of Pediatrics
University of Texas Medical Branch
Galveston, TX 77550

III. Booklets and Videos Explaining PKU to Children and Youth:

You and PKU. (1978, 50 pages: \$5.00)

A booklet for children ages 3 to 8 years old describing PKU and why they need to be on a special diet. Also includes suggestions for parents in teaching their child about PKU.

Marketing Department

University of Wisconsin Press

114 North Murray Street

Madison, WI 53715

All About PKU. (1978, 18 pages: \$1.50)

A cartoon coloring book in which PKU and the diet are explained. This book is appropriate for 4-to 9-year-old children. (Also available in Spanish)

Melanie M. Hunt, R.D.

Metabolic Treatment Center, IDR 528

Children's Hospital Research Foundation

Elland and Bethesda Avenues, Cincinnati, OH 45229

What is PKU? (n.d., 12 pages: single copy free)

Children's coloring and reading book.

PKU Section

Children's Hospital and Health Center

San Diego Regional Center for the Developmentally Disabled

8001 Frost Street, San Diego, CA 92123

Why Is Mary on a Diet? (n.d., 14 pages: single copy free)

Simple explanation of the PKU diet for young children.

Hazel M. Vespa, ACSW

PKU Clinic, The Children's Memorial Hospital

2300 Children's Plaza, Chicago, IL 60614

Hidden from View: PKU and the Teenager (n.d., 20 pages: \$1.00; VHS videotape: \$10.00)

A booklet and video explaining PKU for teenagers. Questions answered include: What is PKU? How did I get PKU? How was I treated for PKU? Why should I think about PKU now? What can I do as a teen?

Pediatric Neurology Metabolic Clinic
Box 54, C1054 Outpatient Building
University Hospital
Ann Arbor, MI 48109

PKU and Teens: Planning Makes it Easier (1987, 29 pages: single copy free)

Designed to help teenagers with PKU learn more about PKU, its treatment and application of dietary needs to situations usually encountered by teenagers.

Illinois Department of Public Health
Division of Family Health
Genetics Diseases Program
535 West Jefferson Street
Springfield, IL 62761

Teenagers and PKU (1988, 18 minute VHS videotape, free on loan)

Companion video to PKU and Teens booklet (above). Discusses issues teens face with respect to continuing the low phe diet; includes a segment on Maternal PKU (two weeks' notice required).

Mead Johnson
Phone 800: 222-9123

IV. Materials Concerning Maternal PKU:

The Young Woman with PKU (1986, 50 pages: \$5.00)

A thorough review of PKU and the required diet, with special emphasis on the problems associated with pregnancy in a woman who has PKU.

PKU Treatment Center, Department of Pediatrics
University of Texas Medical Branch, C-19
Galveston, TX 77550

Understanding Maternal PKU (1990, 12 pages: single copy free)

Booklet written to help women with PKU and their families understand Maternal PKU. Topics include basic information about Maternal PKU, description of PKU, genetics and treatment.

PKU Clinic, Children's Hospital Medical Center
300 Longwood Avenue, Boston, MA 02115

Understanding Mild Hyperphenylalaninemia (1986, 7 pages: single copy free)

Description of the biochemistry, detection and genetics of mild hyperphenylalaninemia

PKU Clinic, Children's Hospital Medical Center
300 Longwood Avenue, Boston, MA 02115

Your Diet for Maternal PKU (1987, 19 pages: single copy free)

Discussion of the need for diet before conception and description of the low phe diet.

(Also available in Spanish)

PKU Clinic, Children's Hospital Medical Center
300 Longwood Avenue, Boston, MA 02115

PKU and Pregnancy (1984, 28 pages: \$3.00)

A guide for women with phenylketonuria and other forms of hyperphenylalaninemia. Topics include the problem of PKU and pregnancy, preparing for pregnancy, the nature of the diet and meal planning.

The Genetic Metabolic Disease Program
The Hospital for Sick Children
555 University Ave., Toronto, Ontario M5G 1X8, CANADA

Maternal Hyperphenylalaninemia (1984, 6 pages: single copy free)

Booklet written for young women with PKU or hyperphenylalaninemia, their families and friends.

PKU Clinic, St. Christopher's Hospital for Children
2603 North 5th Street, Philadelphia, PA 19133

Hidden from View: PKU and the Teenager (n.d., 12 minute VHS videotape: \$10)

A videocassette designed to draw attention to the concerns of PKU and pregnancy.

Pediatric Neurology Metabolic Clinic
Box 54, C1045 Outpatient Building
University Hospital
Ann Arbor, MI 48109

Women with PKU (n.d., 18 pages; single copy free)

Booklet written for young women with PKU, describing the California Maternal PKU project, and answering common questions about PKU and Maternal PKU. A list of all PKU clinics in California is also provided.

California Maternal PKU Project
2125 Berkeley Way, Annex 4, Room 300
Berkeley, CA 94704

PKU, Pregnancy and You (1987, 48 pages; single copy free)

An attractively formatted 5 by 8 inch booklet for young women with PKU with extensive information concerning PKU, its genetic basis, as well as general pregnancy advice, sample diet menus, and answers to commonly asked questions. The list of PKU clinics in California is provided, together with the regional contributing centers of the Maternal PKU Collaborative Study.

California Maternal PKU Project
2125 Berkeley Way, Annex 4, Room 300
Berkeley, CA 94704

V. References for Teachers and Other Professionals:

A Teacher's Guide to PKU. (1985, 13 pages: \$1.00)

Resource book for preschool teachers and school staff; contains PKU basics, NutraSweet warning, suggestions of how teachers can help the child with PKU, questions and answers about PKU, classroom activities.

Maria Nardella, M.A., R.D.
Nutrition Consultant for Crippled Children's Service
Arizona Dept. of Health Services
1740 West Adams (Room 208)
Phoenix, AZ 85007

Games that Teach (1986, 8 pages: \$2.50)

For parents and teachers of preschool and kindergarten classes. Offers nutrition activities appropriate for a group of children including the child with phenylketonuria.

Cristine M. Trahms, M.S., R.D.
PKU Clinic, CDMRC WJ-10
University of Washington, Seattle, WA 98195

PKU for Children: Learning to Measure (1981, 24 pages: \$3.00)

"Lesson" format for parents and teachers. Goal of increased responsibility for self-management is approached through food preparation and accurate measuring.

Cristine M. Trahms, M.S., R.D.
PKU Clinic, CDMRC WJ-10
University of Washington, Seattle, WA 98195

Low Protein Cookery for PKU (1988, 569 pages: \$13.75 plus \$1.50 postage)

Over 450 recipes plus helpful hints for managing the PKU diet.

Marketing Department, University of Wisconsin Press
114 North Murray Street
Madison, WI 53715

Dental Health in Children with PKU (1984, 28 pages: single copy free)

Provides information for medical and dental professionals about the dental health needs of children with PKU.

National Center for Education in Maternal and Child Health
38th and R Streets, N.W.
Washington, D. C. 20057

Screening and Treatment of PKU (1985, slide/sound: 131 slides with a 38 minute audiotape: available for purchase: \$283.80)

Prepared by the staff at the Waisman Center of the University of Wisconsin, Madison, this is a comprehensive introduction to the condition. It includes an explanation of the metabolic basis for PKU, differences between treated and untreated cases, screening of newborns for PKU, and the dietary program needed to prevent damage due to excessive phenylalanine. Designed for parents and older children with PKU as well as professionals.

Distribution Dept., Health Sciences Consortium
201 Silver Cedar Court
Chapel Hill, NC 27514
Phone 919: 942-8731

VI. References in the Professional Literature:

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American Academy of Pediatrics (1965) Committee on the Handicapped Child: statement on treatment of phenylketonuria. *Pediatrics*, 35: 499.

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