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UNIVERSITY OF WASHINGTON

# **Is this Health Information Good for Me? Steps for Finding and Evaluating Health Information on the Web**

**Janet G Schnall, MS, AHIP  
Information Management Librarian  
Health Sciences Libraries  
University of Washington, Seattle, WA  
schnall@u.washington.edu**

# Objectives

- Identify **steps** for finding authoritative PKU and health information on the web
- Describe 3 techniques to **evaluate** health websites

# Trends in Online Health Information

61% of US adults go online for health information.

Most popular topics are diseases and treatments.

Social-networking sites are used only sparingly for health queries and updates.

Fox, Susannah. "The Social Life of Health Information." *Pew Internet & American Life Project*, 2009.

# STEP 1: Start with the best consumer website for health information

National Library of Medicine's



[medlineplus.gov](http://medlineplus.gov)

# MedlinePlus

*medlineplus.gov*

- **#1 SOURCE** for basic quality consumer/patient information
- Includes drug information
- Medical Encyclopedia – full-text with illustrations
- Spanish version
- Preformulated *PubMed* searches
- Interactive tutorials
- Current health news

# MedlinePlus Content

Interactive tutorials:  
**165 total**

ClinicalTrials links on display:  
**640 (today)**

NIH-Seniorhealth  
**43 topics**

OR-Live surgery videos:  
**73**

Announcement listservs:  
**11 daily & weekly**



Health topics (today):  
**800 English**  
**700 Spanish**

3 drug and herbal databases:  
**approx 1700 monographs**

ADAM.com encyclopedia:  
**approx 4000 monographs**

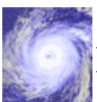
Health news:  
**Approx 15-20 new stories added per day**

Directories:  
**Over 100 directories covering doctors, hospitals, clinics and libraries.**

- Health Topics**  
Start here with over 700 topics on conditions, diseases and wellness
- Drug Information**  
About your prescription and over-the-counter medicines
- Medical Encyclopedia**  
Includes pictures and diagrams
- Dictionary**  
Spellings and definitions of medical words
- News**  
Health News from the past 30 days
- Directories**  
Find doctors, dentists and hospitals
- Other Resources**  
Local libraries, health organizations, international sites and more


- Current Health News**
- ▶ [Mouthguards Essential for Back-to-School Sports](#)
  - ▶ [Newest Breast Cancer Drugs Can Cause Joint Pain in Patients](#)
  - ▶ [Excessively Sleepy? Could Be More Than Poor Sleep](#)
  - ▶ [More news](#)

**Featured Site**



Hurricane Katrina links: [Disasters and Emergency Preparedness](#), [Coping with Disasters](#), [NIH Response](#), [Recovery Information](#)

- In the Spotlight**
- September is Prostate Cancer Awareness Month. Learn more:
- ▶ Go to [Prostate Cancer](#)
  - ▶ [Prostate Cancer Interactive Tutorial](#)
  - ▶ News about [Prostate Cancer](#)

- Interactive Tutorials**  
Over 165 slideshows with sound and pictures
- ClinicalTrials.gov**  
Studies for new drugs and treatments
- NIH SeniorHealth**  
Health information for older adults
- Surgery Videos**  
Videos of surgical procedures
- ▶ What's new on MedlinePlus?  
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  - ▶ Take a [tour](#) of the site

Also called: PKU

Phenylketonuria (PKU) is a genetic disorder in which the body can't process part of a protein called phenylalanine (Phe). Phe is in almost all foods. If the Phe level gets too high, it can damage the brain and cause severe mental retardation. All babies born in U.S. hospitals must now have a [screening](#) test for PKU. This makes it easier to diagnose and treat the problem early.

The best treatment for PKU is a diet of low-protein foods. There are special formulas for newborns. For older children and adults, the diet includes many fruits and vegetables. It also includes some low-protein breads, pastas and cereals. Nutritional formulas provide the vitamins and minerals they can't get from their food.

Babies who get on this special diet soon after they are born develop normally. Many have no symptoms of PKU. It is important that they stay on the diet for the rest of their lives.

National Institute of Child Health and Human Development

Get Phenylketonuria updates by e-mail   [What's this?](#)

## Start Here

- [Genetics Home Reference: Phenylketonuria](#) **NIH** (National Library of Medicine)
- [Phenylketonuria \(PKU\)](#) **NIH** (National Institute of Child Health and Human Development)
- [PKU](#) (March of Dimes Birth Defects Foundation)

Also available in [Spanish](#)

Basics	Learn More	Multimedia & Cool Tools
<ul style="list-style-type: none"><li>• <a href="#">Overviews</a></li></ul>	<ul style="list-style-type: none"><li>• <a href="#">Nutrition</a></li><li>• <a href="#">Disease Management</a></li><li>• <a href="#">Related Issues</a></li></ul>	<ul style="list-style-type: none"><li>• <a href="#">Tutorials</a></li></ul>
Research	Reference Shelf	For You
<ul style="list-style-type: none"><li>• <a href="#">Clinical Trials</a></li><li>• <a href="#">Genetics</a></li><li>• <a href="#">Journal Articles</a></li></ul>	<ul style="list-style-type: none"><li>• <a href="#">Directories</a></li><li>• <a href="#">Organizations</a></li><li>• <a href="#">Law and Policy</a></li></ul>	<ul style="list-style-type: none"><li>• <a href="#">Adults</a></li></ul>



## Encyclopedia

- [Phenylketonuria](#)
- [Serum phenylalanine screening](#)

## Related Topics

- [Genetic Disorders](#)
- [Newborn Screening](#)
- [Brain and Nerves](#)
- [Genetics/Birth Defects](#)
- [Metabolic Problems](#)

## Go Local

Services and providers for **Phenylketonuria** in the U.S.

Select Location   
[Select from list](#)

## National Institutes of Health

## Overviews

- [Phenylketonuria](#) (Mayo Foundation for Medical Education and Research)

## Nutrition

- [At the Table with Family](#) (National PKU News)
- [Is Aspartame Safe for My Child?](#) (Nemours Foundation)
- [Look-Alike Foods: A Solution or a Pandora's Box?](#) (University of Washington PKU Clinic) - PDF
- [What Is the Diet for PKU?](#) (University of Washington PKU Clinic)



## Disease Management

- [How Is PKU Monitored?](#) (University of Washington PKU Clinic)
- [NIH Consensus Panel Recommends Comprehensive Approach to Life Long Care for PKU](#) **NIH** (National Institute of Child Health and Human Development)

## Related Issues

- [Babysitter's Guide to PKU](#) (University of Washington PKU Clinic) - PDF
- [Introduction to Drug Products Containing Phenylalanine](#) (National PKU News)
- [Questions and Answers about PKU](#) (National PKU News)

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## Tutorials

- [What Is Phenylketonuria \(PKU\)?](#) (Dolan DNA Learning Center)

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## Clinical Trials

- [ClinicalTrials.gov: Phenylketonurias](#) **NIH** (National Institutes of Health)

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## Genetics

- [Genetics Home Reference: Phenylketonuria](#) **NIH** (National Library of Medicine)
- [Genetics Home Reference: Tetrahydrobiopterin deficiency](#) **NIH** (National Library of Medicine)
- [Learning about Phenylketonuria \(PKU\)](#) **NIH** (National Human Genome Research Institute)

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## Journal Articles

### References and abstracts from MEDLINE/PubMed (National Library of Medicine)

- [Article: ACOG Committee Opinion no. 449: Maternal phenylketonuria.](#)
- [Article: Changing dietary practices in phenylketonuria.](#)
- [Article: Dietary interventions for phenylketonuria.](#)
- [Phenylketonuria -- see more articles](#)

research on *Phenylketonuria* is the [National Institute of Child Health and Human Development](#)

## Languages

- [Arabic](#) (العربية)
- [Spanish](#) (español)

# Medical Encyclopedia & Dictionary

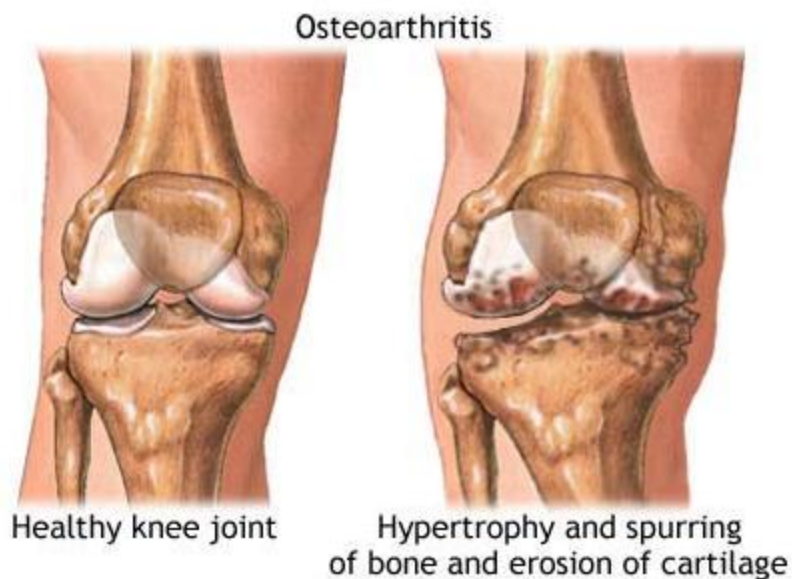
- Encyclopedia includes:
  - 4,000+ entries on diseases, tests, symptoms, injuries and surgeries
  - an extensive library of photographs and illustrations.



 [Home](#) [Health Topics](#) [Drugs & Supplements](#) [Encyclopedia](#) [Dictionary](#) [News](#) [Directories](#) [Other Resources](#)

## Medical Encyclopedia

### Osteoarthritis



ADAM.

Osteoarthritis is a chronic disease of the joint cartilage and bone, often thought to result from "wear and tear" on a joint, although the congenital defects, trauma and metabolic disorders. Joints appear larger, are stiff and painful and usually feel worse the more they

**Update Date:** 7/3/2004

Updated by: Jonathan Harding, MD, CPE, Department of Medicine, University of Massachusetts Medical School, Worcester, MA. Review provided by Ve

# STEP 2: Try other top consumer health websites



# Medical Library Association's Top Ten Most Useful Health Websites

*[mlanet.org/resources/medspeak/topten.html](http://mlanet.org/resources/medspeak/topten.html)*

Cancer.gov

Centers for Disease Control and Prevention  
(CDC) A-Z Topics

FamilyDoctor.org

healthfinder

KidsHealth

Mayo Clinic

MedlinePlus (English | Spanish)

## Quick Guides

- [Living a Healthy Life](#)
- [Students and Campus Life](#)

## Life Stages

- [Children's Health](#)
- [Men's Health](#)
- [Women's Health](#)
- [Senior Health](#)

## Selected Topics

- [Cancer](#)
- [Clinical Trials/Medical Research](#)
- [Diagnostic Tests](#)
- [Drug Information](#)
- [Health Information in Multiple Languages](#)
- [Healthcare Delivery](#)
- [Heart Disease and Stroke](#)
- [Talking to Your Doctor](#)

## Need Help Now?

Immediate assistance is available if you need help dealing with a crisis:

- [Crisis Clinic of King County](#)
- [National Suicide Prevention Lifeline](#)

## Starting Places

Looking for health information? **Your health care provider is your best source of information about your personal health.** Library staff are not qualified to give medical advice, however there are many excellent resources we recommend when researching your health care questions. Here is a selected list of good starting places:

- [MedlinePlus](#)  
Authoritative information from the National Library of Medicine to help answer your health questions.
- [MedlinePlus Guide to Healthy Web Surfing](#)
- [Healthwise® Knowledgebase](#)  
Unbiased information to help consumers make decisions about their health. Access courtesy of Group Health.
- [Centers for Disease Control: A-Z Topics Index](#)
- [MayoClinic.com](#)
- [Merck Manual of Medical Information - Home Edition](#)
- [Merriam-Webster Medical Dictionary](#)

## Find More Information

- [Find a Health Library](#)  
Consumer health libraries providing services to local residents.
- [Search for a library in Washington near you](#)
- [Search for public libraries nationwide](#)
- [Find a Hospital](#)
- [PubMed](#)  
Free resource to locate current biomedical research

## Health Headlines

- [Northern Exposure](#)
- [Smoking May Be in Your Genes](#)
- [Noncardiac Chest Pain May Warrant More Management](#)



[RSS](#)

# Other Authoritative Websites

- Drug and Herbal Sites for Consumers
  - MedlinePlus:  
Drugs, Supplements and Herbal Information  
[www.nlm.nih.gov/medlineplus/druginformation.html](http://www.nlm.nih.gov/medlineplus/druginformation.html)
  - Drug Information Portal  
[druginfo.nlm.nih.gov/drugportal/drugportal.jsp](http://druginfo.nlm.nih.gov/drugportal/drugportal.jsp)
- eTexts
  - Merck Manual of Medical Therapeutics  
[www.merckhomeedition.com](http://www.merckhomeedition.com)
  - eMedicine [www.emedicine.com](http://www.emedicine.com)

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Other drug names: [A](#) [B](#) [C](#) [D](#) [E](#) [F](#) [G](#) [H](#) [I](#) [J](#) [K](#) [L](#) [M](#) [N](#) [O](#) [P](#) [Q](#) [R](#) [S](#) [T](#) [U](#) [V](#) [W](#) [X](#) [Y](#) [Z](#) [0-9](#)

## Sapropterin

(sap" roe ter' in)

 [Printer-friendly version](#)

 [E-mail to a friend](#)

### Contents of this page:

- [Why is this medication prescribed?](#)
- [How should this medicine be used?](#)
- [Other uses for this medicine](#)
- [What special precautions should I follow?](#)
- [What special dietary instructions should I follow?](#)
- [What should I do if I forget a dose?](#)
- [What side effects can this medication cause?](#)
- [What storage conditions are needed for this medicine?](#)
- [In case of emergency/overdose](#)
- [What other information should I know?](#)
- [Brand names](#)

**Why is this medication prescribed?**      [Return to top](#)

Sapropterin is used along with a restricted diet to control blood phenylalanine levels in people who have phenylketonuria (PKU; an inborn condition in which phenylalanine may build up in the blood and causes decreased intelligence and a decreased ability to focus, remember, and

# 105 Herbs and Supplements Monographs in English & Spanish

## Peppermint oil (*Mentha x piperita* L.)

*Natural Standard Bottom Line Monograph, Copyright © 2005 (www.naturalstandard.com). Commercial distribution prohibited. This monograph is intended for informational purposes only, and should not be interpreted as specific medical advice. You should consult with a qualified healthcare provider before making decisions about therapies and/or health conditions.*



*While some complementary and alternative techniques have been studied scientifically, high-quality data regarding safety, effectiveness, and mechanism of action are limited or controversial for most therapies. Whenever possible, it is recommended that practitioners be licensed by a recognized professional organization that adheres to clearly published standards. In addition, before starting a new technique or engaging a practitioner, it is recommended that patients speak with their primary healthcare provider(s). Potential benefits, risks (including financial costs), and alternatives should be carefully considered. The below monograph is designed to provide historical background and an overview of clinically-oriented research, and neither advocates for or against the use of a particular therapy.*

### Related Terms:

- Balm mint, black peppermint, brandy mint, curled mint, Feullis de menthe, Japanese peppermint, Katzenkraut (German), lamb mint, *Mentha arvensis* L. var *piperascens*, menta prima (Italian), *Menthae piperitae aetheroleum* (peppermint oil), *Menthae piperita* var *officinalis*, *Menthae piperitae folium* (peppermint leaf), *Menthe anglaise*, *Menthe poivre*, *Menthe poivree*, *Mentha piperita* var *vulgaris*, Our Lady's mint, pebermynte (Danish), Pfefferminz (German), Porminzen, Schmecker, spearmint (*Mentha spicata* L.), water mint (*Mentha aquatica*), white peppermint, WS(R) 1340.
- **Essential oil constituents:** Cineol, isomenthone, liminene, menthofuran, menthol, menthone, menthyl acetate, terpenoids.
- **Leaf constituents:** Caffeic acid, chlorogenic acid, luteolin, hesperidin, rutin, "volatile" oil.
- **Selected brand names:** Ben-Gay®, Colpermin®, China Maze, Cholaktol, Citaethol, Enteroplant® (contains peppermint and caraway oil), Kiminto, Mentacur, Mentholatum, Mintec, Rhuli Gel®, Robitussin® cough drops, SX Mentha®, Vicks VapoRub®.
- **Combination products:** Absorbine Jr.®, Iberogast®, Listerine®.

## Peppermint oil

## Aceite de menta (*menta piperita*)

*Natural Standard Bottom Line Monograph, Copyright © 2005 (www.naturalstandard.com). Se prohíbe su distribución comercial. Esta monografía tiene la intención de servir para fines informativos únicamente, por lo cual no se debe interpretar como un consejo médico específico. Usted deberá consultar con un proveedor médico calificado antes de tomar decisiones respecto a terapias y/o afecciones de salud.*



*No obstante se han estudiado de forma científica ciertas técnicas complementarias y alternas, para la mayoría de las terapias hay limitación o controversia sobre los datos de alta calidad respecto a la seguridad, eficacia y mecanismo de acción. Se recomienda, al máximo posible, que los practicantes cuenten con licencias expedidas por una organización profesional reconocida que se adhiera a normas claramente publicadas. Además, antes de iniciar una nueva técnica o contratar a un practicante, se recomienda que los pacientes consulten con su(s) proveedor(es) médico(s) principal(es). Se deben considerar atentamente los beneficios y riesgos potenciales (incluye los costos financieros) así como las alternativas. La siguiente monografía está diseñada para ofrecer una historia y un resumen de la investigación con orientación clínica, y la misma ni defiende ni se opone al uso de una terapia en particular.*

### Términos relacionados:

- **Bálsamo de menta, menta negra, menta de brandy, menta crespá, Feullis de menthe, menta japonesa, Katzenkraut (alemán), menta de cordero, menta arvenis, L. var piperascens, menta prima (italiano), Menthae piperitae aetheroleum (aceite de menta) Menthae piperita var officinalis, Menthae piperitae folium (hoja de menta), Menthe anglaise, Menthe poivree, Mentha piperita var vulgaris, Our Lady's mint, pebermynte (danés), Pfefferminz (alemán), Porminzen, Schmecker, hierbabuena (Mentha spicata), menta acuática (Mentha aquatica), menta blanca, WS (R) 1340.**
- **Elementos constituyentes esenciales del aceite:** Cineol, isomentona, limoneno, mentofurano, mentol, mentona, acetato de mentilo, terpenoides.
- **Elementos constituyentes de la hoja:** Ácido cafeico, ácido clorogénico, luteolina, hesperidina, rutin, aceite "volátil".
- **Selección de marcas registradas:** BenGay®, Colpermin®, China Maze, Cholaktol, Citaethol, Enteroplant® (contiene aceite de menta y alcaravea), Kiminto, Mentacur, Mentholatum, Mintec, Rhuli Gel®, Robitussin® cough drops (pastillas para la tos), SX Mentha®, Vicks VapoRub®.

## Evidence [Return to top](#)

*These uses have been tested in humans or animals. Safety and effectiveness have not always been proven. Some of these conditions should be evaluated by a qualified healthcare provider.*

<b>Uses based on scientific evidence</b>	<b>Grade*</b>
<b>Indigestion (non-ulcer dyspepsia)</b> <p>There is preliminary evidence from a small number of controlled trials that a combination of peppermint oil and caraway oil may be beneficial for dyspepsia (heartburn) symptoms. However, most studies have been poorly designed (methodologically weak with small sample sizes, inadequate use of control or placebo groups, unclear descriptions of blinding and randomization, and lack of use of standardized scales for identifying subjects or assessing endpoints). It is not clear which constituent(s) may be beneficial. Nonetheless, the existing evidence does suggest efficacy of this combination. It should be noted that heartburn can actually be a side effect of taking oral peppermint oil, which has been reported by patients in several controlled trials of peppermint oil. Patients with chronic heartburn should be evaluated by a qualified healthcare provider and may be advised to undergo a diagnostic endoscopy prior to initiating any treatment for heartburn.</p>	<b>B</b>
<b>Irritable bowel syndrome (IBS)</b> <p>Multiple randomized controlled trials of peppermint suggest significant improvements in irritable bowel syndrome (IBS) symptoms. Although the mechanism of action is not clear, pre-clinical studies suggest smooth muscle relaxing properties of peppermint (calcium antagonism may play a role). Enteric-coated peppermint preparations are generally recommended. Overall, studies have been brief with small sample sizes and methodological weaknesses (unclear diagnostic criteria, lack of validated measurement scales, unclear blinding and randomization procedures). Well-designed large trials are necessary before a strong recommendation can be made. Future studies should use standardized symptom scales and established diagnostic criteria to classify patients prior to enrollment (such as Rome II Diagnostic Criteria), uniform dosing and standardization, and longer duration.</p>	<b>B</b>
<b>Antispasmodic (gastric spasm)</b> <p>One study reports that peppermint oil solution administered intraluminally can be used as an antispasmodic agent with superior efficacy and fewer side effects than hyoscine-N-butylbromide administered by intramuscular injection during upper endoscopy.</p>	<b>C</b>
<b>Tension headache</b> <p>Application of diluted peppermint oil to the forehead and temples has been tested in people with headache. Studies have not been well conducted, and it is not clear if this is an effective treatment.</p>	<b>C</b>

[Home](#)[News and Features](#)[NLM Resources](#)[NLM Research Resources](#)[Resources by Audience / Class](#)[Other Resources](#)[Home](#) ▶ [Search Results](#)[druginfo.nlm.nih.gov/drugportal/drugportal.jsp](http://druginfo.nlm.nih.gov/drugportal/drugportal.jsp) By Name  By Category[◀ Go back to previous page.](#)

## Search Results

**Drug Name:** Sapropterin [INN] [\[show more names\]](#)

**Search Term:** KUVAN

## Summary

- ▶ [i](#) Summary of drug information (MedlinePlusDrug)
- ▶ [i](#) Summary of consumer health information (MedlinePlusTopics)
- ▶ [i](#) Manufacturers drug label (DailyMed)
- ▶ [i](#) Clinical trials (ClinicalTrials.gov)
- ▶ [i](#) Drug Identification and Image Display (Pillbox *beta*)

## Detailed Summary

- ▶ [i](#) References from scientific journals (Medline/PubMed)
- ▶ [i](#) References from toxicological journals (TOXLINE)
- ▶ [i](#) Biological activities and chemical structures (PubChem)
- ▶ [i](#) Toxicological and chemical resources (ChemIDplus)

## Additional Resources

- ▶ [i](#) Information from the US Food & Drug Administration (Drugs@FDA)
- ▶ [i](#) Search engine for other government resources (USA.gov)

Recent Searches

■ pku

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In This Topic

- Disorders of Amino Acid Metabolism
  - Phenylketonuria (PKU)
    - Symptoms
    - Diagnosis
    - Prognosis
    - Prevention and Treatment
  - Maple Syrup Urine Disease
  - Homocystinuria
  - Tyrosinemia

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## Disorders of Amino Acid Metabolism

Amino acids are the building blocks of proteins and have many functions in the body. Hereditary disorders of amino acid processing can result from defects either in the breakdown of amino acids or in the body's ability to get amino acids into cells. Because these disorders cause symptoms early in life, newborns are routinely screened for several common ones. In the United States, newborns are commonly screened for phenylketonuria, maple syrup urine disease, homocystinuria, tyrosinemia, and a number of other inherited disorders, although screening varies from state to state.

### PHENYLKETONURIA (PKU)

*Phenylketonuria occurs in infants born without the ability to normally break down an amino acid called phenylalanine. This disorder causes phenylalanine, which is toxic to the brain, to build up in the blood.*

- Phenylketonuria is caused by lack of the enzyme needed to convert phenylalanine to tyrosine.
- Symptoms include intellectual disability, seizures, nausea, vomiting, an eczema-like rash, and a mousy body odor.
- The diagnosis is based on a blood test.
- A strict phenylalanine-restricted diet allows for normal growth and development.

Phenylketonuria (PKU) is a disorder that causes a buildup of the amino acid phenylalanine, which is an essential amino acid that cannot be synthesized in the body but is present in food. Excess phenylalanine is normally converted to tyrosine, another amino acid, and eliminated from the body. Without the enzyme that converts it to tyrosine, phenylalanine builds up in the blood and is toxic to the brain, causing intellectual disability.

#### Symptoms

Newborns with PKU rarely have symptoms right away, although sometimes they are sleepy or eat poorly. If not treated, affected infants progressively develop intellectual disability over the first few years of life, and it eventually becomes severe. Other



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Pronunciations

amniocentesis

chorionic villus sampling

cobalamin

cystinuria

dialysis



# eMedicine

*emedicine.medscape.com*

**eMedicine**  
from WebMD



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eMedicine Specialties > Pediatrics: Genetics and Metabolic Disease > Metabolic Diseases

## Phenylketonuria

**Author: Georgianne L. Arnold, MD**, Director of Inherited Metabolic Disorders Clinic, Department of Pediatrics and Genetics, Associate Professor, University of Rochester School of Medicine and Dentistry  
[Contributor Information and Disclosures](#)

Updated: Feb 13, 2009

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### Introduction

#### Background

Phenylketonuria (PKU) is an inborn error of protein metabolism that results from an impaired ability to metabolize the essential amino acid phenylalanine. Classic phenylketonuria is present when plasma phenylalanine levels exceed 20 mg/dL (1200 mmol/L) without treatment. Lesser degrees of elevation of plasma phenylalanine are considered in [Hyperphenylalaninemia](#). Elevated phenylalanine negatively impact developmental function, and individuals with classic phenylketonuria almost always have mental retardation unless levels are controlled through dietary treatment. In the U.S. and many other countries, phenylketonuria is detected through newborn screening, and treated individuals have normal intelligence.

#### Pathophysiology

Most individuals with phenylketonuria have a deficiency of the enzyme phenylalanine hydroxylase. Phenylalanine hydroxylase deficiency is inherited in an autosomal recessive manner.

#### Overview

#### Differential Diagnoses & Workup

#### Treatment & Medication

#### Follow-up

#### Multimedia

### References

1. Yannicelli S, Ryan A. Improvements in behaviour and physical manifestations in previously untreated adults with phenylketonuria using a phenylalanine-restricted diet: a national survey. *J Inherit Metab Dis*. 1995;18(2):131-4. [[Medline](#)].
2. Burton BK, Grange DK, Milanowski A, et al. The response of patients with phenylketonuria and elevated serum phenylalanine to treatment with oral sapropterin dihydrochloride (6R-tetrahydrobiopterin): a phase II, multicentre, open-label, screening study. *J Inherit Metab Dis*. Oct 2007;30(5):700-7. [[Medline](#)].
3. Schindeler S, Ghosh-Jerath S, Thompson S, et al. The effects of large neutral amino acid supplements in PKU: an MRS and neuropsychological study. *Mol Genet Metab*. May 2007;91(1):48-54. [[Medline](#)].
4. Sarkissian CN, Gamez A, Wang L, et al. Preclinical evaluation of multiple species of PEGylated recombinant phenylalanine ammonia lyase for the treatment of phenylketonuria. *Proc Natl Acad Sci U S A*. Dec 30 2008;105(52):20894-9. [[Medline](#)].

STEP 3: For more technical health  
information in journal articles,  
search *PubMed*



# PubMed

*pubmed.gov*

- Includes citations to biomedical journal articles from the 1940's to date
- Indexes 5,200 biomedical journals
- 75-80% of citations have abstracts
- Updated 5x/week
- Links to full-text articles

## Limits

**Can easily limit your search to:**  
**Age groups**  
**Human or Animal studies**  
**Language**  
**Review articles**  
**Only items with links to full text**

### Dates

Published in the Last: Any date

### Type of Article

- Meta-Analysis
- Practice Guideline
- Randomized Controlled Trial
- Review

### Species

- Humans
- Animals

### Subsets

#### Journal Groups

- Core clinical journals
- Dental journals
- Nursing journals

### Languages

- English
- French
- German
- Italian
- Japanese

### Gender

- Male
- Female

### Ages

- All Infant: birth-23 months
- All Child: 0-18 years
- All Adult: 19+ years
- Newborn: birth-1 month

# PubMed Results

## [Sapropterin: a new therapeutic agent for phenylketonuria.](#)

Hegge KA, Horning KK, Peitz GJ, Hegge K.

Ann Pharmacother. 2009 Sep;43(9):1466-73. Epub 2009 Aug 4. Review.

PMID: 19654333 [PubMed - indexed for MEDLINE]

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## [Efficacy of sapropterin dihydrochloride in increasing phenylalanine tolerance in children with phenylketonuria: a phase III, randomized, double-blind, placebo-controlled study.](#)

Trefz FK, Burton BK, Longo N, Casanova MM, Gruskin DJ, Dorenbaum A, Kakkis ED, Crombez EA, Grange DK, Harmatz P, Lipson MH, Milanowski A, Randolph LM, Vockley J, Whitley CB, Wolff JA, Bechuk J, Christ-Schmidt H, Hennermann JB; Sapropterin Study Group.

J Pediatr. 2009 May;154(5):700-7. Epub 2009 Mar 4.

PMID: 19261295 [PubMed - indexed for MEDLINE]

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## [Optimizing the use of sapropterin \(BH\(4\)\) in the management of phenylketonuria.](#)

Blau N, Bélanger-Quintana A, Demirkol M, Feillet F, Giovannini M, MacDonald A, Trefz FK, van Spronsen FJ.

Mol Genet Metab. 2009 Apr;96(4):158-63. Epub 2009 Feb 8. Review.

PMID: 19208488 [PubMed - indexed for MEDLINE]

[Related citations](#)

## [Safety and efficacy of 22 weeks of treatment with sapropterin dihydrochloride in patients with phenylketonuria.](#)

Lee P, Treacy EP, Crombez E, Wasserstein M, Waber L, Wolff J, Wendel U, Dorenbaum A, Bechuk J, Christ-Schmidt H, Seashore M, Giovannini M, Burton BK, Morris AA; Sapropterin Research Group.

Am J Med Genet A. 2008 Nov 15;146A(22):2851-9.

PMID: 18932221 [PubMed - indexed for MEDLINE]

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## Efficacy of sapropterin dihydrochloride in increasing phenylalanine tolerance in children with phenylketonuria: a phase III, randomized, double-blind, placebo-controlled study.

Trefz FK, Burton BK, Longo N, Casanova MM, Gruskin DJ, Dorenbaum A, Kakkis ED, Crombez EA, Grange DK, Harmatz P, Lipson MH, Milanowski A, Randolph LM, Vockley J, Whitley CB, Wolff JA, Bebhuk J, Christ-Schmidt H, Hennermann JB; Sapropterin Study Group.

▶ Collaborators (16)

Klinik für Kinder-und Jugendmedizin Reutlingen, Klinikum am Steinenberg, Reutlingen, Germany. [ftrefz@ramedis.de](mailto:ftrefz@ramedis.de)

### Abstract

**OBJECTIVE:** To evaluate the ability of sapropterin dihydrochloride (pharmaceutical preparation of tetrahydrobiopterin) to increase phenylalanine (Phe) tolerance while maintaining adequate blood Phe control in 4- to 12-year-old children with phenylketonuria (PKU). **STUDY DESIGN:** This international, double-blind, randomized, placebo-controlled study screened for sapropterin response among 90 enrolled subjects in Part 1. In Part 2, 46 responsive subjects with PKU were randomized (3:1) to sapropterin, 20 mg/kg/d, or placebo for 10 weeks while continuing on a Phe-restricted diet. After 3 weeks, a dietary Phe supplement was added every 2 weeks if Phe control was adequate. **RESULTS:** The mean (+/-SD) Phe supplement tolerated by the sapropterin group had increased significantly from the pretreatment amount (0 mg/kg/d) to 20.9 (+/-15.4) mg/kg/d ( $P < .001$ ) at the last visit at which subjects had adequate blood Phe control ( $<360$  micromol/L), up to week 10. Over the 10-week period, the placebo group tolerated only an additional 2.9 (+/-4.0) mg/kg/d Phe supplement; the mean difference from the sapropterin group (+/-SE) was 17.7 +/- 4.5 mg/kg/d ( $P < .001$ ). No severe or serious related adverse events were observed. **CONCLUSIONS:** Sapropterin is effective in increasing Phe tolerance while maintaining blood Phe control and has an acceptable safety profile in this population of children with PKU.

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## [Phenylketonuria: MedlinePlus Medical Encyclopedia](#)

Mar 23, 2010 - **PKU** is a treatable disease. **Treatment** involves a diet that is extremely low in phenylalanine, particularly when the child is growing. ...

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## [What we know that could influence future treatment of phenylketonuria](#)

CN Sarkissian, A Gamez, CR Scriver - *Journal of inherited metabolic ...*, 2009 - Springer  
Summary **Phenylketonuria (PKU)**, a Mendelian auto- somal recessive phenotype (OMIM 261600), is an inborn error of metabolism that can result in impaired postnatal cognitive development. The phenotypic out- come is multifactorial in origin, based both in nature, ...

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## [New approaches to the treatment of phenylketonuria](#)

O Freitas, C Izumi, MG Lara, LJ Greene - *Nutrition Reviews*, 2009 - [interscience.wiley.com](#)

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FJ van Spronsen, GM Enns - *Molecular Genetics and Metabolism*, 2010 - Elsevier

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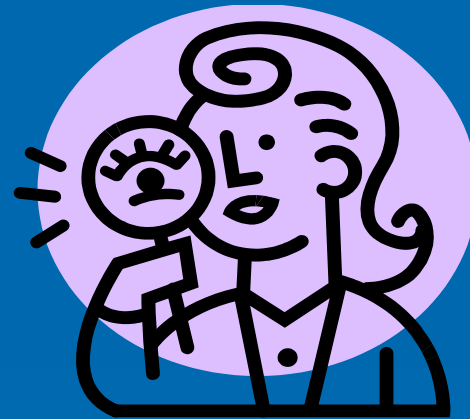
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# Criteria for Evaluating Web Sites

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- Authority
- Accuracy
- Objectivity
- Currency
- Coverage
- Design



# Authority



- Is the author clearly identified?
- Are the author's credentials listed?
- Is there an institutional affiliation?
- Does the author have standing in the field?
- Can you easily contact the author for clarification of information?

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# What is PKU?

by Dr. Tracy L. Beck, Ph.D. ?

Have you ever looked at the label on a diet soda can and seen the warning: **Phenylketonurics - contains phenylalanine**? I personally know of several people who refuse to drink diet soda because the word "phenylketonurics" sounds too scary! What does "phenylketonurics" mean? Is it something you can catch from diet soda?

Well, let me take the opportunity to answer these questions. "Phenylketonurics" is NOT something you can catch from diet soda! This long, scary-sounding word is included in a warning at the end of the ingredients list on some products, but it is NOT an ingredient these products. "Phenylketonurics" is the term used to refer to people that have the metabolic disorder Phenylketonuria, or PKU for short. So - kind of like how the word "diabetics" refers to people that have "diabetes", "phenylketonurics" refers to the people that have "phenylketonuria". I myself, am one of these phenylketonurics and the warning on diet soda cans is included merely to inform people like me that the product contains the synthetic chemical ASPARTAME . People that have the disorder PKU cannot consume any product that contains aspartame, for reasons which I will elaborate on below.

Two additional questions that people often ask when they learn of the true meaning of the warning label on diet soda are - Do I have PKU? How do I know that I don't have PKU? Well, if you have reached an age where you have the desire to drink diet soda and you still have the mental capacity to question whether or not you have PKU, the odds are that you don't have the disorder.

Phenylketonuria is a genetic metabolic disorder that results when the PKU gene is inherited from both parents. When babies are born in the United States, a heel stick blood test is done to test for various disorders - PKU is one of the disorders that is tested for. It is very rare, in the US an average of 1 in 15,000 babies are born with the disorder (this number varies from region to region). If a baby is tested positive for PKU at birth, a second test is done a few days later. Out of 500 babies which do test positive for PKU on the first test, on average only 100 actually have the disorder.



So, what is PKU? What happens to people that have it?

I want to note that I am NOT a medical professional (my Ph.D. is in Astronomy, not medicine), and you should never make important decisions about your health without the guidance of a medical specialist. However, if you think you may be experiencing effects of aspartame poisoning, I recommend an easy test - avoid all aspartame containing products for 1

# What is PKU?

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One will notice that many products clearly state whether or not they contain phenylalanines. You can see such labels on sodas, as well as on packaged nuts and cheese in most cases. As well as avoiding protein rich foods, most starch-laden foods like potatoes and pasta have to be avoided as well, so the diet is limited. To give the body the necessary protein it needs, protein formulas have been specifically developed for those with PKU so they can grow and develop normally.

When women with PKU become pregnant, the presence of PKU in the fetal environment is linked to birth defects of varied types. It is particularly important that pregnant women with PKU seek the advice of a high-risk [obstetrician](#). Most important is a strict adherence to a PKU diet during the pregnancy to prevent birth defects in the fetus.

Since the diet of the person affected by PKU is restrictive, there are many companies that now manufacture PKU foods that are very low in or completely absent of phenylalanine. These products, like egg substitutes, ice cream substitutes, and various soups or spaghetti sauces, as well as low protein breads and pastas, can help the person with PKU. The cost of such food can be expensive, particularly for the more fancy substitutions.

With the help of a dietitian, a person with PKU can live a long and healthy life. His or her food choices may be limited, but generally PKU will cause no other symptoms or result in complications to the person's health.

Written by Tricia Ellis-Christensen

PKU or [phenylketonuria](#) is a condition that affects about 1 in 15,000 people. Those with PKU lack the enzyme phenylalanine hydroxylase, which allows the body to turn ingested phenylalanine into the [amino acid tyrosine](#). Tyrosine helps the body synthesize proteins, and where it is not present, the results can be quite devastating.

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## [PKU - Phenylketonuria](#)

Forums, Help, Support, Information SHS International PKU Community [www.shs-nutrition.com](http://www.shs-nutrition.com)

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Newborns are routinely checked for PKU, because if they have the condition, which is inherited recessively, they must be on a special diet which is very low in phenylalanine. Phenylalanine unfortunately occurs in most proteins, like eggs, nuts, meat, and breast or dairy milk. Phenylalanine is also present in most infant formulas. If the child with PKU eats these foods, he or she will develop irreversible [mental retardation](#), hence the mandatory testing.

The testing for PKU is fairly simple, though a bit hard to watch for new moms. The newborn's heel is pricked, then a small amount of blood is squeezed out and sent to a lab for analysis. The lab results usually come back within a few days, since following dietary restrictions are vital for the newborn, and the child or adult with PKU.

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# About wiseGEEK

*Who is behind wiseGEEK?* It's one of the most common questions we receive. We are a team of researchers, writers and editors dedicated to providing clear and concise answers to common questions. Currently, there are over 100 active contributors; you can read some of their profiles below.

Tricia has been part of the wiseGEEK team since 2006, and in that time she's contributed more than 2,500 articles to the site. As a graduate in literature from [Sonoma State University](#), her passion in life is reading, reading, and then more reading, and she is extraordinarily grumpy when she runs out of books. Writing, as the natural extension of reading, has served Tricia well as a wiseGEEK contributor. She's been able to explore more of her interests, researching topics including medicine, art, film, history, politics, ethics, and religion. Tricia remains fascinated by the connectivity between different areas of thought and knowledge, and hopes her work helps to create bridges of understanding between reader, writer and topic. Tricia lives in Northern California with her beloved husband of over 15 years and her two sons, and is continuing work on her first novel. Some of her articles include: [What is Fundamentalism?](#), [Who are Beren and Luthien?](#), and [Why is a Rabbit's Foot Considered Lucky?](#)



"wiseGEEK gives me an opportunity to view the world from so many different perspectives and explore such diverse avenues of thought. I love finding the answers to our readers' questions and find it wonderful to have a job that allows me to constantly learn new things!"



## What is PKU?

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*Phenylketonuria (PKU) is a genetic inborn error of metabolism that is detectable during the first days of life with appropriate blood testing (newborn screening). The absence or deficiency of an enzyme that is responsible for processing the essential amino acid phenylalanine characterizes PKU. With normal enzymatic activity, phenylalanine is converted to another amino acid (tyrosine), which is then utilized by the body. However, when the phenylalanine hydroxylase enzyme is absent or deficient, phenylalanine abnormally accumulates in the blood and is toxic to brain tissue.*

*Without treatment, most infants with PKU develop mental retardation. Those with untreated PKU may also develop additional neurologic symptoms.*

*To prevent mental retardation, treatment consists of a carefully controlled, phe-restricted diet begun during the first days or weeks of life. Most experts suggest that a phe-restricted diet should be lifelong. A carefully maintained diet can prevent mental retardation as well as neurological, behavioral and dermatological problems. It is generally believed that keeping blood phenylalanine levels in the range of 2-6mg/dl is the safest, especially in infancy and early childhood. Frequent blood monitoring should be done to achieve this goal.*

*PKU is inherited as an autosomal recessive trait. In other words, two people who conceive a child must both be carriers of the gene in order for there to be a chance that their infant will have PKU. When both carriers conceive a child, there is a one in four or 25% chance for each pregnancy that the baby will have PKU. It is estimated that PKU occurs in one in 15,000 newborns in the United States. The incidence varies in other parts of the world.*



# Cristine M. Trahms Program for Phenylketonuria • Seattle

- WHO WE ARE
- WHAT WE OFFER
- CLINIC SCHEDULE
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- HOW TO FIND US

## WHO WE ARE

The Cristine M. Trahms Program for Phenylketonuria at the University of Washington is made up of a team of experienced health care professionals. Together, this team provides services to people with PKU and their families in the state of Washington.

The program is primarily funded by the [Washington State Newborn Screening Program](#). The State Genetics Program provides families with subsidized formula (paid for by their medical insurance). The programs are closely coordinated to ensure follow-up and appropriate management of individuals with PKU.

### PKU Clinic Staff

- C. Ronald Scott, MD, Clinic Director
- Anne Leavitt, MD, Clinic Co-Direcor
- Cristine Trahms, MS, RD, Program Director
- Janie Heffernan, MS, RD, Nutritionist
- Beth Ogata, MS, RD, Nutritionist
- Janet Garretson, MSW, Social Worker
- Stefanie Uhrich, MSc, CGC, Genetic Counselor
- Vicki Frasher, Patient Care Coordinator
- Steve Sulzbacher, PhD, Consulting Psychologist
- Anne Hay, PhD, Psychologist



# Accuracy

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Phenylketonuria (PKU) is a genetic disorder that causes the amino acid phenylalanine to build up in the blood. This results in severe and irreversible mental retardation, especially in infants, if not treated early.

There is little information available on the incidence of PKU. In the United States, research reports:

no date

- PKU occurs in 1 out of every 13,500 to 19,000 births.
- A higher incidence in whites and **Native Americans**.
- A lower incidence in blacks, Hispanics, and Asians.

no source for references

### Cause and Incidence

In PKU, the child lacks or has very low levels of the enzyme phenylalanine hydroxylase (PAH). PAH is necessary to convert phenylalanine into another amino acid, tyrosine. If phenylalanine is not converted, it builds up in the bloodstream and disturbs the metabolism, resulting in mental retardation. Phenylalanine is a component of protein, so when protein is consumed, phenylalanine builds up in those people with PKU.

At birth, babies with PKU have no signs or symptoms of the disorder because the mother's body filters out phenylalanine for her unborn baby before birth. Symptoms appear after the baby starts consuming protein (from either breast milk or formula), and the resulting phenylalanine builds up in his or her system, usually by a few months of age. If PKU is not treated, it usually causes progressively severe mental retardation and other problems affecting the nervous system. The baby will not be able to regain the lost mental ability.

PKU is an autosomal recessive disease. This means that to have the disease, a person must inherit a gene from both parents. If a person inherits the gene from only one parent, he or she is a carrier. This means that the person carries the gene for PKU, but does not have the disease.

### Treatment

In the United States, babies are routinely tested for PKU shortly after birth so that early diagnosis and treatment are possible and so that injury to the baby's brain is usually prevented. Treatment for PKU consists of a special diet containing reduced amounts of protein (which contains phenylalanine).

If you think your new baby has symptoms of phenylketonuria, talk with your health professional about whether the baby was tested adequately. Early diagnosis and treatment for PKU can prevent mental retardation. See the Exams and Tests section of this topic for more information on testing.

## What Does it Mean when Your Baby Has PKU?



Published August 08, 2006 by:

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Phenylketonuria is a metabolic disorder, where the individual is unable to metabolize a protein that is known as phenylalanine. The buildup of the phenylalanine within the blood stream can actually interfere with the

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development of the brain and cause serious retardation to the baby. Although this disorder is not very common you are probably wondering what causes it and who is susceptible to this type of disorder? In order for your baby to be born with PKU both parents must carry the recessive gene. If both parents do carry the trait, than your baby will have a one in five chance of being born with PKU. In order for the doctor to be able to determine if your baby has PKU they will need to perform a blood test within the first few days of birth.

If your baby is diagnosed with PKU the doctor will put

Renee writes in her spare time for websites, and parenting magazines. She enjoys reading and writing articles on education, music, sports, fishing, swimming, healthcare, parenting, and childcare. With a background in education, she focuses most of her articles on children, and various teaching methods.



and meat. This diet will usually have to be started earlier than the treatment begins the better the prognosis the more difficult it will be to follow that you follow all of the doctor's directions you need in order to advance in life normally

Some parents are not aware that they already had one child and they have not

yourself and your husband tested. Remember even if you both carry the recessive gene it does not mean that all of your children will get PKU. It is also important to keep in mind that if your baby is diagnosed with this disorder it does not mean that they will have problems for the rest of their life. What happens to then as a result of this disorder is really of to you and the doctors. To learn more about this disorder you can speak to your doctor and find out if you need to be concerned about PKU.

H.J. Breck,  
M.D. PKU  
Clinic  
Director  
(Guest)  
03/04/2007

The article on PKU by rene has several very serious inaccuracies and misleading information and wrong facts. Please take it off your website and rewrite it with the true facts about PKU. This disorder is treated for life not just "8 or 9 years" as stated. The risk of PKU is one in four not "1 in 5" as your article stated. The parents of "one child without PKU" do not need to have genetic testing! Did you mean "one child with PKU"? It is a disservice to families looking on the internet to be given such a poorly researched and written article about a metabolic disorder. It is very damaging and dangerous to propagate wrong information.

# Morbidity and Mortality Weekly Report (MMWR)

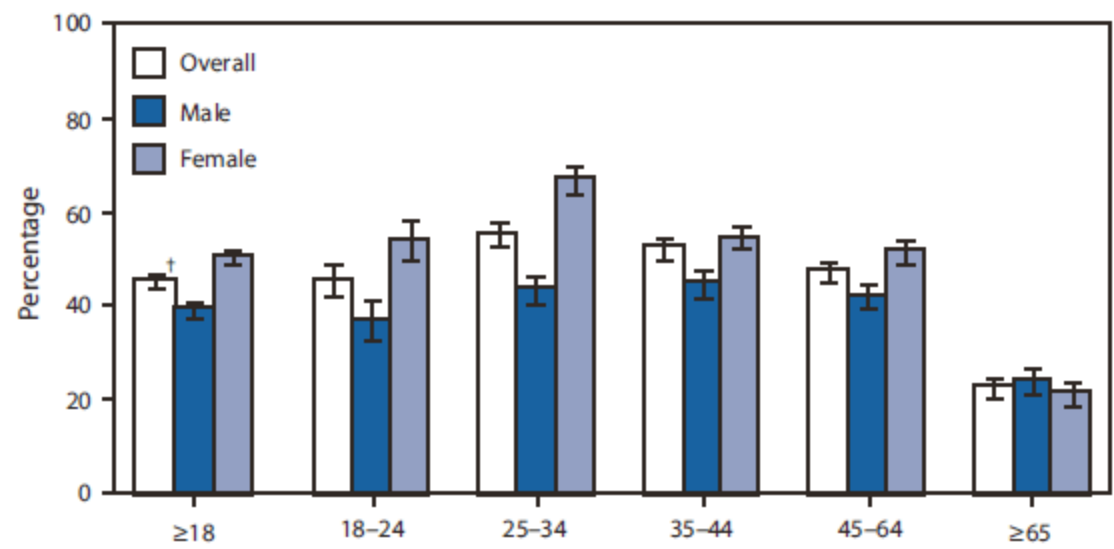
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Text size: **S** M L



## QuickStats: Percentage of Adults Aged >18 Years Who Looked Up Health Information on the Internet, by Age Group and Sex\*--- National Health Interview Survey, United States, January--September 2009

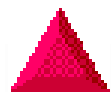
*Weekly*  
April 23, 2010 / 59(15);461



# Objectivity

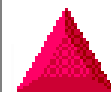
- Is the purpose of the site clearly stated?
- Is the information impartially presented?
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The complete reference center and  
information source for MELATONIN



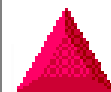
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- [What](#) exactly is melatonin?
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### Miscellaneous

- Our [Code of Ethics](#)
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E-mail Address:  
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SALE!	Regularly ...	Now
Melatonin, Regular Release	\$9.95...	\$6.95!
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DHEA, Sustained Release	\$14.95...	\$7.95!
<i>ArthRight!</i> , Osteoarthritis Remedy	\$29.95...	\$19.95!

[ArthRight!](#) by Worldwide Labs(tm)

900 mg Tablets, 60 per bottle, Qty 1-5: **\$19.95** each, Qty 6-11: \$18.95 each

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Fish Oil and Fish Oil Benefits

fishoil on 22 Apr 2009 07:50 am

## Fish Oil: Countering the Bad Effects of PKU

**TAKE THE OMEGA-3 CHALLENGE**

And.. Find out why  
Triple Strength Omega-3  
Is the best fish oil

Try A Bottle On Us!  
**FREE TRIAL**



Loss of ability to break down an amino acid called phenylalanine is called PKU. It is a rare condition which results when both parents pass down this disease through their genes. The baby acquires phenylketonuria or PKU through autosomal recessive trait.

The babies with PKU are deficient in the enzyme, phenylalanine hydroxylase, which is vital for breaking down phenylalanine. If not broken down, this

no author  
no refs



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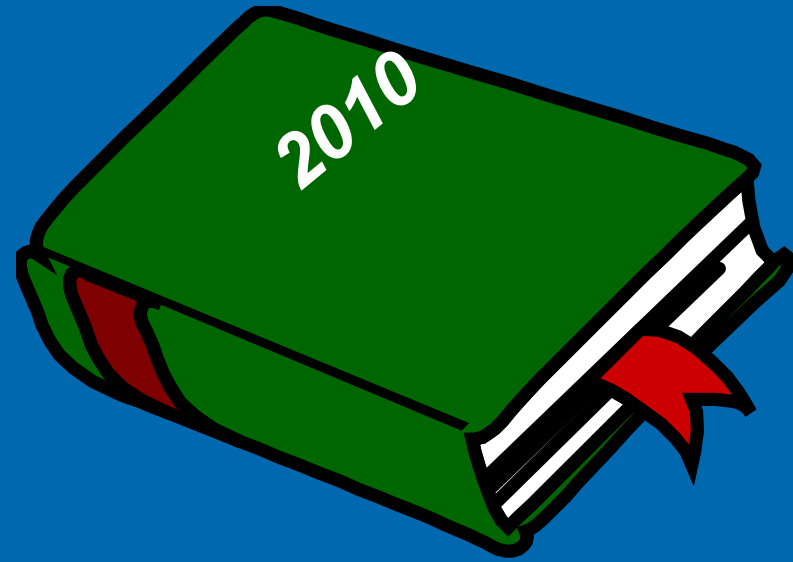
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# Coverage

- Does the site satisfy the needs of its intended audience?
- Is the subject coverage comprehensive?

# Currency



- Is the site current?
- Was the site updated recently?
- Are the links kept up-to-date?

# Phenylketonuria

## Inherited metabolic disorder

From [Mary Kugler, R.N.](#), former About.com Guide *Updated: March 29, 2009*

About.com Health's Disease and Condition content is reviewed by our [Medical Review Board](#)

See More About: [newborn screening](#) [metabolic disorders](#) [genetic testing](#)

Phenylketonuria (fen-ul-keetone-YU-ree-ah, or PKU) is an inherited metabolic disorder. In PKU, the body cannot completely break down the protein phenylalanine because a necessary enzyme, phenylalanine hydroxylase, is deficient. Because of this, phenylalanine builds up in the body's cells and causes nervous system and brain damage.



### About.com **Medical Review Board**

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[Kate Grossman, MD](#)

#### Medical Director

A board-certified internal medicine physician, Dr. Kate Grossman joined About.com in September 2006 as Medical Director. She oversees About.com's medical review board, comprised of physicians from prominent medical institutions who review content for medical accuracy. In addition, she serves as the in-house medical consultant for the shaping of About.com Health and its properties.

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Phenylketonuria is a treatable by a simple blood test. In the [newborn babies be tested for](#) Kingdom, Canada, Australia, t

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## Phenylketonuria (PKU)

### What is phenylketonuria (PKU)?

Phenylketonuria (pronounced fee-nill-key-toe-NURR-ee-uh) or PKU is an inherited disorder of metabolism that can cause intellectual and developmental disabilities (IDDs) if not treated. In PKU, the body can't process a portion of the protein called phenylalanine (Phe), which is in almost all foods. If the Phe level gets too high, the brain can become damaged.

All babies born in U.S. hospitals are now routinely tested for PKU soon after birth, making it easier to diagnose and treat them early.

### What are the symptoms of PKU?

Children with untreated PKU may appear normal at birth. By age three to six months, they begin to lose interest in their surroundings. By age one year, they are developmentally delayed and their skin has less pigmentation than someone without the condition. If Phe is not restricted in the diet, those with PKU develop severe IDDs.

### What are the treatments for PKU?

The most effective treatment for PKU is a special diet of foods that help control

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Before you consider trying the new PKU drug, Kuvan, please click [here](#) for important new information.

**Recent Website Updates** - January 2010

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E-mail comments or questions to Virginia Schuett: [schuett@pkunews.org](mailto:schuett@pkunews.org)

# ABOUT National PKU News

## What is the National PKU News Organization?

*By Virginia Schuett, MS, RD, Director and Editor*

*6869 Woodlawn Ave. NE #116*

*Seattle, WA 98115-5469*

*Tel: 206-525-8140*

*Fax: 206-525-5023*

*Email: [schuett@pkunews.org](mailto:schuett@pkunews.org)*

## Professional and Personal Tidbits

National PKU News is a non-profit organization located in Seattle, Washington. It is dedicated to providing up-to-date, accurate news and information to families and professionals dealing with phenylketonuria. I started the organization in the Spring of 1989, after 16 years as nutritionist for a large PKU clinic in Madison, Wisconsin. The organization relies on generous donations from subscribers to continue thriving (all donations are tax-deductible for US residents).

National PKU News now has over 2000 subscribers who receive the organization's newsletter. Most subscribers are from the US, but an increasing number are from other countries (35 countries are now

National PKU News has an excellent Board of Directors:

Cris Trahms, MS, RD from the Seattle PKU Clinic, Seattle, WA and  
Dorothy Corry, PKU parent of a young adult, from Seattle, WA.

## What is PKU?

*By Virginia E. Schuett, MS, RD, Director, National PKU News*

PKU (phenylketonuria), in its "classic" form, is a rare, inherited metabolic disease that results in mental retardation and other neurological problems when treatment is not started within the first few weeks of life. When treatment is begun early and well-maintained, affected children can expect normal development and a normal life span. (Not all elevations of blood phenylalanine require treatment; any child with a level less than 6 mg/dl does not need to be on a special diet and is not risk for mental retardation.) Until recently, the only treatment was a very strict diet that limits the amino acid, phenylalanine. Now, however, there is a drug called Kuvan that can help in some cases.

### **The Enzyme Deficiency**

The disease arises from the absence of a single enzyme called phenylalanine hydroxylase. This enzyme normally converts the essential amino acid, phenylalanine, to another amino acid, tyrosine. Failure of the conversion to take place results in a buildup of phenylalanine. Through a mechanism that is not well understood, the excess phenylalanine is toxic to the central nervous system and causes the severe problems normally associated with PKU. Not every child has the same degree of enzyme deficiency and thus the treatment for each child has to be individualized. The nature of the treatment for an individual child must be determined by an experienced PKU treatment program.

### **Genetics**


PKU is carried through an "autosomal recessive" gene. This means that two people who conceive a child must both be "silent carriers" of the gene

Last update: February 2008

National PKU News: [www.pkunews.org](http://www.pkunews.org)

E-mail: [schuett@pkunews.org](mailto:schuett@pkunews.org)

# Design

- Is the site clearly organized?
  - Is there an internal search engine?
  - Is the site user friendly?
  - Can this site be accessed reliably?
- 
- A decorative graphic in the bottom right corner of the slide, consisting of several concentric circles of varying sizes and colors (light blue, white, and dark blue) that resemble ripples in water.

# Analyze the Website address

- edu
- org
- com
- Gov
- net

*how information  
is transmitted*

*sub directory*

*http://www.cdc.gov/nip/child.htm*

*name of host computer*

*filename*

The URL (Uniform Resource Locator) includes the name of the host computer which can indicate the purpose of the web site.

# Guides for Evaluating Health Information

- Criteria for Evaluating Web Resources  
*[healthlinks.washington.edu/howto/navigating/criteria.pdf](http://healthlinks.washington.edu/howto/navigating/criteria.pdf)*
- MedlinePlus Guide to Healthy Web Surfing  
*[www.nlm.nih.gov/medlineplus/healthywebsurfing.html](http://www.nlm.nih.gov/medlineplus/healthywebsurfing.html)*
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*[www.mlanet.org/resources/userguide.html](http://www.mlanet.org/resources/userguide.html)*



# Privacy Policy

- Read Privacy Policy
- Does site ask for personal information?
- How will your information be used?
- Are you comfortable with how site will use your information?
- Consider a separate email account

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- ADULTS INTRODUCTION ▶
- PARENTS INTRODUCTION ▶
- ABOUT PKU.COM** ▼
- ABOUT PKU ▶
- EATING WELL WITH PKU ▶
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## About PKU.com

PKU.com is your online resource for all things PKU. Here you can learn about PKU, find out about new research, and discover ways to manage PKU, including diet, medication and other resources.

You can also be part of the PKUiverse, a safe and private online community where you can get ideas, share your thoughts, ask questions and make friends:

- Customize your own PKU home page
- Join chat, blogs, and other programs
- Upload your thoughts, ideas, recipes, and videos
- And much more.

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# PKU and you

pku.com

## Taking charge of your life, diet and health

You may have thought that you could quit worrying about Phe when you grew up. But today, we know that high Phe causes problems for teens and adults, not just children.<sup>1</sup>

Many studies show that high Phe is bad for the brain throughout life. Too much Phe can make it harder to think. It can make you more moody. High Phe makes it hard to focus and pay attention and can even slow down your ability to react.<sup>1,3,4,5</sup>



1. Mitchell J, Scriver CR. Phenylalanine hydroxylase deficiency. Bethesda (MD): National Library of Medicine (US), National Center for Biotechnology Information; Gene Reviews. 2007 March. Available from: <http://www.genetests.org/query?dz=pku>. Accessed February 3, 2009.
2. Scriver CR, Kaufman S. Hyperphenylalaninemia: phenylalanine hydroxylase deficiency. In: Scriver CR, Beaudet AR, Sly W, Valle D, eds. *The Metabolic and Molecular Bases of Inherited Disease*. 8th ed. New York, NY: McGraw-Hill; 2001:1667-1724.
3. Waisbren SE, Noel K, Fahrback K, et al. Phenylalanine blood levels and clinical outcomes in phenylketonuria: a systematic literature review and meta-analysis. *Mol Genet Metab*. 2007;92:63-70.
4. Moyle JJ, Fox AM, Arthur M, Bynevelt M, Burnett JR. Meta-analysis of neuropsychological symptoms of adolescent and adults with PKU. *Neuropsychol Rev*. 2007;17:91-101.
5. VanZutphen KH, Packman W, Sporri L, et al. Executive functioning in children and adolescents with phenylketonuria. *Clin Genet*. 2007;72:13-18.

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- Will disclose aggregate info
- Emails, chats, message boards, etc. not private
- If follow links, check PP
- Can Opt-out of emails and newsletters

### MedicineNet Privacy Policy

WebMD understands how important the privacy of personal information is to our users. This Privacy Policy will tell you what information we collect about you and about your use of WebMD and its services. It will explain the choices you have about how your personal information is used and how we protect that information. We urge you to read this Privacy Policy carefully.

We have organized this Privacy Policy under the following topics:

[PART 1: Our Accreditation and Privacy Seals](#)

[PART 2: About This Privacy Policy and Using Our Site and Tools](#)

[PART 3: Non-Personal Information We Collect About You](#)

- Cookies
- Web Beacons
- Third Parties Collecting Non-Personal Information on WebMD's Behalf

[PART 4: Personally Identifiable Information We Collect About You](#)

- Newsletters & Emails to You
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- Message Boards and other Public Forums
- Website Registration and Interactive Tools on WebMD
- Subscription Services
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- Concept Unique Identifiers
- Aggregate Data
- Secure Message Center
- Interactive Tools on WebMD Health Manager

[PART 6: Information Collected by Third Parties Not Acting on Behalf of WebMD](#)

[PART 7: Disclosure of Your Information](#)

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- Disclosure to or by Co-branded Channel Partners
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[PART 8: How WebMD Handles Privacy and Security Internally](#)

[PART 9: Updating Your Information and Contacting WebMD](#)

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- Removing your Personally Identifiable Information
- Updating Your Personal Health Information in WebMD Health Manager
- Limitations on Removing or Changing Information
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[PART 11: Glossary](#)

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When you visit our website, our web server automatically recognizes only the Internet domain and IP address from which you accessed our website. This information does not result in the identification of your personal e-mail address or other personal information.

In addition, we gather information regarding the volume and timing of access to our website by collecting information on the date, time and website pages accessed by visitors to the website. We do this so we can improve the content of our website and this information is not shared with other organizations. Again, only aggregate information is collected and individual visitors' personal information is not identified.

If you choose to share personal information with us - by sending us a message, or filling out an electronic form with personal information - we will use the information only for the purposes you authorized or in support of university sponsored programs - we will not disclose the information to other third parties or government agencies, unless required to do so by state or federal law or in support of university sponsored programs.

Enter search term here

SEARCH

Diseases and Conditions

Symptoms

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More than 3,300 physicians, scientists and researchers from Mayo Clinic share their expertise to empower you to manage your health.

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### Tests and Procedures

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Information and tools for a healthy lifestyle.

## Diseases and Conditions

Find a disease by its first letter

- A
- B
- C
- D
- E
- F
- G
- H
- I
- J
- K
- L
- M
- N
- O
- P
- Q
- R
- S
- T
- U
- V
- W
- X
- Y
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- #



### Raw milk debate heats up

Fans of raw milk say it tastes better and has health benefits. Public health experts counter that raw milk poses a health threat.

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- Advertising and sponsorship policy
- Advertising and sponsorship opportunities

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Scott C. Litin, M.D.

Editor in chief of "Mayo Clinic Family Health Book," Third Edition, Dr. Scott Litin is a practicing general internist at Mayo Clinic, Rochester, and a professor at Mayo Clinic College of Medicine.

A distinguished practitioner, lecturer and teacher, Dr. Litin has served in numerous leadership positions at Mayo Clinic and nationally. He's a recipient of many awards, including the prestigious Henry S. Plummer Distinguished Physician Award and the Distinguished Clinician Award from Mayo Clinic. The American College of Physicians has recognized him as a Master of the College.

Dr. Litin has authored numerous medical journal articles and serves in editorial capacities for several Mayo Clinic publications. He's actively involved in continuing education programs for practicing physicians and is frequently an invited speaker at medical gatherings.

**Authority**

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# Phenylketonuria (PKU)

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## Treatments and drugs

By Mayo Clinic staff

Authority

The main phenylketonuria treatment is a low-phenylalanine diet. Doctors used to believe that a low-phenylalanine diet was necessary in adolescence, but today it's not necessary for most people.

A safe amount of phenylalanine is determined by a doctor. Doctors use charts and blood levels to determine a safe amount. You may need to monitor PKU levels as you grow, especially during growth spurts and pregnancy. The amount of phenylalanine in your diet affects your body processes, but not your intelligence.

### Which foods to avoid

Because the amount of phenylalanine adults can safely eat is so low, it's

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Accuracy

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### Phenylketonuria and Other Metabolic Diseases: Clinical, Genetic and Newborn Screening Aspects

Dr. Michael Marble

#### 1. Phenylketonuria (PKU)

##### What is PKU?

Phenylketonuria (PKU) is a genetic condition associated with abnormally high levels of phenylalanine in the body. Elevated phenylalanine leads to increased levels of phenylketones in the blood which are excreted in the urine, thus the name phenylketonuria. In order to understand PKU, it is necessary to understand some basic concepts of metabolism, which is the process by which compounds from our diet are chemically changed and used by our body to carry out the basic functions of life.

Metabolic processes occur along multiple steps called pathways. Each step in the pathway is catalyzed by a specific enzyme. Enzymes are special proteins which act as catalysts to induce chemical changes in other substances in our body. These enzymes are manufactured by our bodies in response to instructions contained in our genes.

Phenylalanine is one of the amino acids, which are important nutrients that we get from our diets. Amino acids are used by the body to make protein which is essential for most physiological processes. Amino acids also serve as precursors to be converted by enzymes into other important compounds for the brain and other organs.

The metabolic pathway we are dealing with in PKU is the conversion of phenylalanine into another amino acid, tyrosine. The importance of this pathway is that it removes excess phenylalanine and it enables the production of sufficient tyrosine. Tyrosine is important for the production of neurotransmitters that function in the brain. The enzyme phenylalanine hydroxylase (PAH) is responsible for enabling the phenylalanine to tyrosine conversion to take place.



First, Second and Third grades; Frisco School; Lions, LA; St. John the Baptist Parish; School was abolished in 1930's

#### Contact Information:

Dr. Mike Marble  
LSU Dept. of Pediatrics  
Children's Hospital  
200 Henry Clay Ave.  
New Orleans, La 70118  
Tel: 504- 896- 9254  
E- mail: [mmarbl1@lsuhsc.edu](mailto:mmarbl1@lsuhsc.edu)

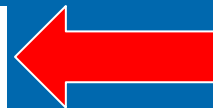


About the Photos



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**Dr. Michael Marble** is a clinical and metabolic geneticist and a faculty member of the Louisiana State University Health Sciences Center. He directs the Metabolic/ PKU clinic located at Children's Hospital of New Orleans. He is co- medical director of the Children's Hospital Metabolic Laboratory and is chairperson of the newborn screening sub- committee.



**Author of PKU article**

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Let the reader and viewer beware.



Use *Steps for Finding Health Information on the Web*

Remember *MedlinePlus, PubMed, My Health* and more

*Evaluate* what you find on the web!



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