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## **PKU (PHENYLKETONURIA) COVERAGE OF TESTING AND TREATMENT**

### **Policy**

VCHCP covers the testing for and treatment of phenylketonuria (PKU) for members of any age when deemed medically necessary to avert the development of serious physical or mental disabilities or to promote normal development or function as a consequence of PKU. VCHCP covers special enteral formulas and specially formulated food, when such products are used in place of normal products and to the extent that their cost exceeds the cost of a normal diet.

### **Coverage of Special Formulas**

Coverage includes special formulas and diet powders for use in the dietary management of members with PKU. Their restriction in phenylalanine content differentiates these metabolites from other products. These enteral products are available in liquid form or in a diet powder intended to be mixed with semisolid food, water or other beverages. Certain PKU formulas are on the Plan's Drug Formulary. The names of these products can be obtained by contacting the Customer Service Department. Other PKU formulas and diet powders are covered if prior approval is obtained from the plan.

### **Coverage of Special Food Products**

Coverage of special food products is limited to food products that are specially formulated to have less than one gram of protein per serving. These special food products are used in place of normal food products. Not included are foods that are naturally low in protein or available to the general population in a grocery store. Examples of special food products are apple fiber or low protein flour used in baking, and low protein spaghetti, flour, crackers, cookies, pretzels, jello and tomato sauce. Plan prior authorization is required. For each food item, the member must submit unit price, Nutrition Facts (including protein grams per serving), quantity requested and expected monthly consumption. The member is also encouraged to submit comparison costs of normal foods. After coverage is authorized, the member is required to pay out of pocket and seek reimbursement from the plan. Not covered is the cost of cook books, recipes, subscriptions, catalogues or cooking equipment (such as bread machines).

## **PKU (PHENYLKETONURIA), COVERAGE OF TESTING AND TREATMENT**

### **Coverage Criteria**

The following criteria are used by the plan in making coverage determinations. The plan's Medical Director will evaluate all but item a (which is an administrative calculation) below:

- a) The special formula or special food product is covered to the extent the cost exceeds the cost of a normal diet. The initial monthly usage is estimated and then actual monthly usage is tracked and reported to VCHCP at least quarterly. The unit cost of the special formula or food product is compared to the unit cost of standard formula or a standard food product available to general

public in a grocery store. The difference in unit cost and the units consumed (or estimated to be consumed) determines the amount of coverage authorized. (This amount will be reimbursed to the member for special food products or converted to units authorized for special enteral formula.)

- b) The prescribed diet must be deemed medically necessary to avert the development of serious physical or mental disabilities or to promote normal development or function as a consequence of PKU. A blood phenylalanine of more than 6 mg/dl may signify that a special diet is needed.
- c) Special food products that are prescribed must be consistent with the best practices of qualified health professionals with expertise germane to, and experience in the treatment and care of PKU.
- d) Members taking special formulas must be under the direct and continuing supervision of a health care professional, who shall manage the member's diet in consultation with a plan-referred endocrinologist. A licensed physician must prescribe the special diet, usually this physician will be a plan-referred endocrinologist.

#### **Healthy Families Program (HFP)**

Requests for treatment for PKU for HFP members will be referred to California Children's Services for a determination of medical eligibility. Dietary recommendations made by a CCS-referred endocrinologist will be considered as having been made by a VCHCP-referred endocrinologist.

**A. Attachments:** None

#### **B. References:**

*SB148 (1999), Health and Safety Code §1374.56*

*Mead Johnson, Product Description for Health Care Professionals, Dietary management of Individuals with Metabolic Disorders*

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

#### **C. History:**

Reviewers: Pamela K. Lindeman Chief Operations Officer, Richard O. Ashby, MD  
Reviewed/Revised: Faustine Dela Cruz, RN & Albert Reeves, MD; Date: November 7, 2011  
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