

Letter of Medical Necessity

Date:

TO: _____ [Flex Spending Account Administrator]

FROM: _____ [Physician Name]

_____ [Physician Address]

_____ [Physician Office Phone #]

_____ [Physician Office Fax #]

SUBJECT: Request for Flex Spending Account reimbursement for Flavis[®] dietary foods

This letter of medical necessity is regarding the nutrition management of _____ [Name], __/__/__ [Date of Birth]. This patient has an inborn error of metabolism, a genetic disorder, known as **Phenylketonuria (PKU) (ICD 10: E70.0)** and is being followed in the genetic/metabolic clinic of [CLINIC NAME]. This letter is to request coverage of **Dr. Schar USA's FLAVIS[®] dietary foods**, a medical food/formula(s) used for the treatment of PKU.

Phenylketonuria (PKU) is an autosomal recessive inborn error of metabolism resulting from a deficiency of phenylalanine hydroxylase (PAH; 612349). PAH is an enzyme that catalyzes the hydroxylation of phenylalanine to tyrosine, the rate-limiting step in phenylalanine catabolism. PKU is part of the newborn screening profile due to the devastating effects if undiagnosed or untreated. An untreated child with PKU will suffer irreversible brain damage as well as severe and progressive neurological disorders. Normal development and growth are possible if an infant with PKU is treated appropriately immediately upon diagnosis.

The three primary goals of treatment are:

1. To maintain the blood phenylalanine at a level to prevent toxicity to the brain
2. To ensure that energy intake is sufficient and sustained throughout the day to prevent fasting and breaking down of body proteins
3. To ensure that the individual's overall nutritional requirements are met, allowing for normal growth and development and the avoidance of nutritional deficiencies

The standard of care for PKU remains as lifelong compliance with a phenylalanine restricted diet and the consumption of special medical foods/formulas prescribed by a licensed physician. This dietary treatment requires specialized management that is supervised by a Geneticist and implemented by a registered dietitian specially trained in the nutrition management of inborn errors of metabolism. It requires frequent blood testing as well as close nutrition follow-up with the registered dietitian and medical team. The recommended treatment ranges of blood levels for individuals with phenylketonuria are generally:

- Phenylalanine: 2 to 6mg/dL (120 and 360 μ mol/L)

Letter of Medical Necessity

There is a strong correlation between cognitive function and maintenance of blood phenylalanine levels in this treatment range. Elevated blood phenylalanine in patients has been associated with behavior and learning problems that can reverse when the blood levels return to the treatment range. If dietary management is not sustained, neurological deterioration, phobias, difficulty in concentration, loss of impulse control, and decrease in IQ points can occur. Continuation of dietary management for life is recommended for all patients with PKU. This recommendation is based on strong evidence indicating there is a decline in average IQ and developmental difficulties in school or work performance after diet discontinuation.

The specialized medical nutrition management for PKU involves:

- A phenylalanine-restricted diet that excludes all foods high in protein (e.g. meat, poultry, fish, eggs, cheese, dairy, nuts, and legumes) and markedly restricts all grains, including rice, breads, and pastas
- Special low-protein modified food products (which are not available in grocery stores)
- Sufficient calorie intake that is evenly distributed throughout the day to prevent catabolism
- Medical foods/formulas that are low or free of phenylalanine that come from powdered or liquid formulas, bars, puddings, gels, or tablets to provide 80-90% of protein intake

The term medical food/formula, is defined in section 5(b) of the Orphan Drug Act {21 U.S.C. 360ee (b) (3)}: a “food which is formulated to be consumed or administered enterally under the supervision of a physician and which is intended for the specific dietary management of a disease or condition for which distinctive nutritional requirements, based on recognized scientific principles, are established by medical evaluation.”

Currently, this patient is prescribed **Dr. Schar USA FLAVIS[®] foods**, which *is a/are* medical food/formula(s) used to manage PKU. Medical nutrition therapy must also provide a sufficient and balanced intake of other nutrients to avoid nutritional deficiencies. Nutrition therapy of PKU without the use of medical food/formula is not possible, because it would cause severe protein malnutrition, calorie deprivation, vitamin and mineral deficiency, failure-to-thrive, and, ultimately, death.

FLAVIS brand products are available directly from the manufacturer, Dr Schar USA.

_____ [Name] requires _____ [# OF GRAMS]
grams of protein equivalents per day from medical food/formula; therefore, we have prescribed
requires _____ [AMOUNT OF PRODUCT] per day to meet the protein
requirements for PKU.

We appreciate your attention to this request for **FLAVIS[®]** medical food/formula(s),
_____ [AMOUNT OF PRODUCT] to be covered by their current medical
insurance.

Please do not hesitate to contact us if you have any questions.

Letter of Medical Necessity

Sincerely,

_____ [Physician name, M.D.]

_____ [Physician's credentials, contact info, clinic name]

_____ [Dietitian name, RD, LDN]

Cc: _____ [Parents' names]

References:

1. National Institute of Health (n.d.) Brief history of newborn screening. Retrieved March 27, 2017, from <https://www.nichd.nih.gov/health/topics/newborn/conditioninfo/pages/history.aspx>
2. Brown, C.S., & Lichter-Konecki, U. (2016). Phenylketonuria (PKU): a problem solved? *Mol Genet Metab Rep.* 6, 8-12.
3. Bernstein, L.E., Rohr, F., & Helm, J.R. (Eds.). (2015). *Nutrition Management of Inherited Metabolic Disorders, Lessons from Metabolic University*. New York: Springer International Publishing.
4. Boyer, S.W., Barclay, L.J., & Burrage, L.C. (2015). Inherited Metabolic Disorders: Aspects of Chronic Nutrition Management. *Nutr Clin Pract.* Aug; 30(4):502-10.
5. Hafid, N.A., & Christodoulou, J. (2015). Phenylketonuria: a review of current and future treatments. *Translational Pediatrics*, 4(4), 304–317.
6. Singh, R.H., Rohr, F., Frazier, D., Cunningham, A., Mofidi, S., Ogata, B., et al. (2014). Recommendations for the nutritional management of phenylalanine hydroxylase deficiency. *Gen Med* 16(2):121-131.
7. Camp, K.M., Lloyd-Puryear, M.A., & Huntington, K.L. (2012). Nutritional treatment for inborn errors of metabolism: indications, regulations, and availability of medical foods and dietary supplements using phenylketonuria as an example. *Mol Genet Metab.* September; 107(1-2): 3–9.
8. Acosta, P.B., and Matalon, K.M. (2010). Chapter 5 Nutrition management of patients with inherited disorders of aromatic amino acid metabolism. In PB Acosta (Eds.), *Nutrition Management of Patients with Inherited Metabolic Disorders* (pp. 119-153). Sudbury, Massachusetts: Jones and Bartlett Publishers.
9. Williams, R.A., Mamotte, C.D.S., Burnett, J.R. (2008). Phenylketonuria: an inborn error of phenylalanine metabolism. *Clin Biochem Rev.* Vol 29: 31-41.