



MASSACHUSETTS

Blue Cross Blue Shield of Massachusetts is an independent licensee of the Blue Cross and Blue Shield Association

Medical & Pharmacy Medical Policy Special Foods

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Policy Number: 304

BCBSA Reference Number: None

Related Policies

None

Policy

Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity

Note: All requests for indications listed and not listed on the medical policy guidelines may be submitted to BCBSMA Clinical Pharmacy Operations by completing the Prior Authorization Form on the last page of this document. Physicians may also call BCBSMA Pharmacy Operations department at (800)366-7778 to request a prior authorization/formulary exception verbally.

Prior Authorization Information

| | | |
|--|---|---|
| <input checked="" type="checkbox"/> Prior Authorization <input type="checkbox"/> Step Therapy <input type="checkbox"/> Quality Care Dosing | | Pharmacy Operations: Tel: 1-800-366-7778 Fax: 1-800-583-6289 Policy last updated 5/2024 |
| Pharmacy (Rx) or Medical (MED) benefit coverage | <input checked="" type="checkbox"/> Rx <input checked="" type="checkbox"/> MED | To request for coverage: Physicians may call, fax, or mail the attached form (Formulary Exception/Prior Authorization form) to the address below. |
| Policy applies to Commercial Members: <ul style="list-style-type: none"> • Managed Care (HMO and POS), • PPO and Indemnity • MEDEX with Rx plan • Managed Major Medical with Custom BCBSMA Formulary • Comprehensive Managed Major Medical with Custom BCBSMA Formulary • Managed Blue for Seniors with Custom BCBSMA Formulary | | Blue Cross Blue Shield of Massachusetts Pharmacy Operations Department 25 Technology Place Hingham, MA 02043 Individual Consideration: Policy for requests that do not meet clinical criteria of this policy, see section labeled Individual Consideration |

Inborn errors of metabolism

As required by Massachusetts state law, specialized formula appropriate to the condition may be **covered with no age limit** for metabolic diseases for patients with the following diagnoses. Documentation required includes the diagnosis.

- Tyrosinemia
- Homocystinuria
- Maple syrup urine disease
- Propionic acidemia
- Methylmalonic acidemia
- Urea cycle disorders
- Phenylketonuria (PKU)*
- Other organic and amino acidemias.

*PKU benefits are provided for infants and children as well as for the protection of unborn babies of women who have PKU.

Malabsorption

As required by Massachusetts state law, Specialized formula appropriate to the condition may be covered for patients **with no age limit** with the following diagnoses:

- Crohn's disease
- Ulcerative colitis
- Gastrointestinal dysmotility
- Gastroesophageal reflux (GERD)
- Chronic intestinal pseudo-obstruction.

Documentation required to demonstrate malabsorption includes the diagnosis, any other pertinent clinical Information AND evidence of growth failure (as defined below), including a copy of the growth chart.

Growth failure is defined as:

- Decreased growth velocity resulting in a decrease of two major growth percentiles (e.g., from 50th percentile to 10th percentile) on a standard growth chart, **OR**
- Weight that is less than 75% of the median weight for age and gender.

Formula intolerance

Non-hypoallergenic formula and Partially Hydrolyzed Protein formulas may be **covered up to 12 months of age** for infants with evidence of growth failure (as defined above), including a copy of the growth chart and documented symptoms of formula intolerance.

Hypoallergenic formulas such as Extensively Hydrolyzed Protein formulas and Amino Acid formulas may be **covered up to 12 months of age** for these infants with formula intolerance if the patient fails a trial of Partially Hydrolyzed Protein formula.

Protein Allergy

Hypoallergenic formulas such as Extensively Hydrolyzed Protein formulas and Amino Acid formulas may be **covered up to 12 months of age** for these infants with a protein allergy as defined below

Covered conditions related to formula intolerance:

- A. IgE-Mediated protein allergy, such as Allergic Eosinophilic Esophagitis. Documentation required includes the diagnosis, symptoms and signs (e.g., urticaria, angioedema, vomiting or an acute flare of atopic dermatitis) and any other pertinent clinical information, **OR**
- B. Symptoms of protein allergy Not Due to IgE-Mediated Conditions. Documentation required includes symptoms of formula intolerance, any other pertinent clinical information.

Prematurity

Formulas formulated for premature infants may be covered for those infants born at less than 36 weeks gestation. The formulas may be **covered for up to 6 months of age**. Documentation required includes gestational age.

Cystic Fibrosis

Specialized formulas may be **covered with no age limit** for patients with Cystic Fibrosis. Documentation required includes the diagnosis.

We may cover **Relizorb**® for the treatment of hydrolyzing fats in enteral formula when all of the following criteria are met:

- Patient has Diagnosis of Cystic Fibrosis, **AND**
- Age is equal to or greater than two (2) years of age, **AND**
- Receiving overnight tube feedings, **AND**
- Not using a formula with insoluble fats

Growth Failure Unrelated to Formula Intolerance, Milk Protein Allergy, Prematurity, Inborn Errors of Metabolism, or Malabsorption Due to the Diagnoses Listed Above

Formulas supplemented with fats or carbohydrates may be **covered up to 12 months of age** for patients who:

- Meet the definition of growth failure above, **AND**
- Have had a complete medical evaluation to diagnose and treat, or rule out, potential causes of the growth failure such as gastrointestinal congenital defects, heart failure secondary to congenital heart disease, infection, adenoidal hypertrophy, diabetes mellitus, immunodeficiency, and neurologic disease. Documentation required includes the diagnosis, a description of the relevant evaluation, any other pertinent clinical information, and a copy of the growth chart.

We may cover enteral formulae prescribed by a licensed practitioner for use in enteral feeding tubes (N-G tube, N-E feeding tubes, G-tubes, J-tubes) with a written physician's order. Feedings must exceed 750 kilocalories a day (for adult patients) to be considered medically necessary.⁴ Please note there are no kilocalories minimums in pediatric patients. Tube feedings are required under the following circumstances:

- An anatomic or structural problem that prevents food from reaching the stomach, for example a tumor or stricture of the esophagus or stomach, or neck cancer.
- A neurological problem that results in swallowing and chewing problems that may lead to aspiration.

We may cover use of an FDA-approved infusion pump to administer the enteral feedings in the following circumstances:

- For patients with dumping syndrome who cannot tolerate bolus feedings, or
- For use in J-tube (jejunostomy) feedings, or
- For patients with inflammatory bowel disease who require small amounts of slow, continuous feedings, or
- For patients who, through surgery or congenital abnormality, do not have a stomach and who require slow, continuous feedings.

We may cover ketogenic diet for children with seizures, refractory to or intolerant of multiple anti-epileptic drugs.^{10,16,17,19}

PKU Guidelines:³

- Treatment of neonates born with PKU should begin 7-10 days after birth.¹⁴
- All infants with blood phenylalanine levels over 10 mg/dL measured while eating a normal protein diet (2-3 grams protein/kg/day), and in whom other amino acids levels, such as tyrosine, are low or normal.

- The PKU enteral formula should be enriched with tyrosine and provide 2-3 grams protein/kg/day. It should be taken as evenly as possible throughout the day.
- Blood phenylalanine levels should be monitored weekly¹⁴ during periods of rapid growth, fluctuating blood levels, or when food intake is unpredictable. In older children and adults, this monitoring can occur 1-2 times per month. The ideal time for this blood test is 2 hours after eating.
- Diet, including special formula intake, is modified to achieve optimal blood levels.
- Optimal blood phenylalanine levels:
 - under age 10 2-6 mg/dL
 - over age 10 2-10 mg/dL
 - women trying to conceive¹⁴ 2-6 m/dL
 - pregnant women¹⁴ 2-6 m/dL
- Women who wish to have children should optimize their levels 2-3 months before conception,¹⁴ and continue close nutritional monitoring during pregnancy.

Standard, non-hydrolyzed, non-supplemented, non-elemental milk and soy-based formulas are not covered as these preparations are considered food and not considered treatment for a medical condition.

According to our subscriber certificates, we do not cover any over-the-counter nutritional items. However, we do adhere to the Massachusetts State mandate.

- We do not cover special medical formulas or non-prescription enteral formulae when used for other conditions not listed above.
- We do not cover blenderized baby food or regular store-bought food for use with an enteral feeding system.
- We do not cover use of over-the-counter foods or prescription foods when store-bought food meets the nutritional needs of the patient.
- We do not cover food for the ketogenic diet, since this diet uses regularly available foods.

Individual Consideration

All our medical policies are written for the majority of people with a given condition. Each policy is based on medical science. For many of our medical policies, each individual's unique clinical circumstances may be considered in light of current scientific literature. Physicians may send relevant clinical information for individual patients for consideration to:

Blue Cross Blue Shield of Massachusetts
 Pharmacy Operations Department
 25 Technology Place
 Hingham, MA 02043
 Tel: 1-800-366-7778
 Fax: 1-800-583-6289

CPT Codes / HCPCS Codes / ICD Codes

Inclusion or exclusion of a code does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage as it applies to an individual member. Providers should report all services using the most up-to-date industry-standard procedure, revenue, and diagnosis codes, including modifiers where applicable.

The following codes are included below for informational purposes only; this is not an all-inclusive list.

The above medical necessity criteria MUST be met for the following codes to be covered for Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity:

- If a member has a pharmacy benefit, enteral formulae must be processed through the pharmacy benefit.
- Participating pharmacies under contract with our Pharmacy Benefit Manager (PBM), will bill all outpatient retail drug claims through the pharmacy on-line system.
- A participating home infusion therapy provider may bill for enteral formulae for members who do not have a pharmacy benefit.

HCPCS Codes

| HCPCS codes: | Code Description |
|--------------|--|
| B4034 | Enteral feeding supply kit; syringe fed, per day, includes but not limited to feeding/flushing syringe, administration set tubing, dressings, tape |
| B4035 | Enteral feeding supply kit; pump fed, per day, includes but not limited to feeding/flushing syringe, administration set tubing, dressings, tape |
| B4036 | Enteral feeding supply kit; gravity fed, per day, includes but not limited to feeding/flushing syringe, administration set tubing, dressings, tape |
| B4081 | Nasogastric tubing with stylet |
| B4082 | Nasogastric tubing without stylet |
| B4083 | Stomach tube - Levine type |
| B4087 | Gastrostomy/jejunostomy tube, standard, any material, any type, each |
| B4088 | Gastrostomy/jejunostomy tube, low-profile, any material, any type, each |
| B4102 | Enteral formula, for adults, used to replace fluids and electrolytes (e.g., clear liquids), 500 ml = 1 unit |
| B4103 | Enteral formula, for pediatrics, used to replace fluids and electrolytes (e.g., clear liquids), 500 ml = 1 unit |
| B4104 | Additive for enteral formula (e.g., fiber) |
| B4105 | In-line cartridge containing digestive enzyme(s) for enteral feeding, each |
| B4148 | Enteral feeding supply kit; elastomeric control fed, per day, includes but not limited to feeding/flushing syringe, administration set tubing, dressings, tape |
| B4149 | Enteral formula, manufactured blenderized natural foods with intact nutrients, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit |
| B4150 | Enteral formula, nutritionally complete with intact nutrients, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit (use for Enrich, Ensure, Ensure HN, Ensure Powder, Isocal, Lonalac Powder, Meritene, Meritene Powder, Osmolite, Osmolite HN, Portagen Powder, Sustacal, Renu, Sustagen Powder, Travasorb) |
| B4152 | Enteral formula, nutritionally complete, calorically dense (equal to or greater than 1.5 kcal/ml) with intact nutrients, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit (use for Magnacal, Isocal HCN, Sustacal HC, Ensure Plus, Ensure Plus HN) |
| B4153 | Enteral formula, nutritionally complete, hydrolyzed proteins (amino acids and peptide chain), includes fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit (Criticare HN, Vivonex t.e.n. (use for Total Enteral Nutrition), Vivonex HN, Vital (Vital HN), Travasorb HN, Isotein HN, Precision HN, Precision Isotonic) |
| B4154 | Enteral formula, nutritionally complete, for special metabolic needs, excludes inherited disease of metabolism, includes altered composition of proteins, fats, carbohydrates, vitamins and/or minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit (use for Hepatic-aid, Travasorb Hepatic, Travasorb MCT, Travasorb Renal, Traum-aid, Tramacal, Aminaid) |
| B4155 | Enteral formula, nutritionally incomplete/modular nutrients, includes specific nutrients, carbohydrates (e.g., glucose polymers), proteins/amino acids (e.g., glutamine, arginine), fat (e.g., medium chain triglycerides) or combination, administered through |

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| | an enteral feeding tube, 100 calories = 1 unit (use for Propac, Gerval Protein, Promix, Casec, Moducal, Controlyte, Polycose Liquid or Powder, Sumacal, Microlipids, MCT Oil, Nutri-source) |
| B4157 | Enteral formula, nutritionally complete, for special metabolic needs for inherited disease of metabolism, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit |
| B4158 | Enteral formula, for pediatrics, nutritionally complete with intact nutrients, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber and/or iron, administered through an enteral feeding tube, 100 calories = 1 unit |
| B4159 | Enteral formula, for pediatrics, nutritionally complete soy based with intact nutrients, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber and/or iron, administered through an enteral feeding tube, 100 calories = 1 unit |
| B4160 | Enteral formula, for pediatrics, nutritionally complete calorically dense (equal to or greater than 0.7 kcal/ml) with intact nutrients, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit |
| B4161 | Enteral formula, for pediatrics, hydrolyzed/amino acids and peptide chain proteins, includes fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit |
| B4162 | Enteral formula, for pediatrics, special metabolic needs for inherited disease of metabolism, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit |

Policy History

| Date | Action |
|----------------|---|
| 5/2024 | Updated pediatric age indication for Relizorb [®] . |
| 10/2023 | Clarified coding information. |
| 7/2023 | Reformatted Policy. |
| 6/2023 | Clarified this is both a Medical and Pharmacy policy and which sections are State mandated. |
| 4/2023 | Removed reference to both Retail benefit and requirement to have Pharmacy Carved-in. |
| 7/2022 | Updated Requirements and added Milk Protein to the policy. |
| 2/2021 | Updated to add coverage for Relizorb [®] to the policy. |
| 7/2018 | Clarified coding information. |
| 10/2017 | Clarified coding information. |
| 6/2017 | Updated address for Pharmacy Operations. |
| 4/2015 | Reviewed - Medical Policy Group - Allergy and ENT/Otolaryngology. Changes to Formula intolerance section were recommended and adopted. |
| 7/2014 | Updated Coding section with ICD10 procedure and diagnosis codes, effective 10/2015. |
| 8/2012 | Updated 8/1/2012 as follows: BCBSMA will now cover formula for children up to one year of age prescribed for malabsorption or intolerance only when criteria for growth failure are met. Formula prescribed for prematurity will be covered up to six months of age. Formula prescriptions for other conditions (with the exception of cystic fibrosis) for members older than age one year must also meet criteria for growth failure. |
| 11/2011-4/2012 | Medical policy ICD 10 remediation: Formatting, editing and coding updates. No changes to policy statements. |
| 1/2012 | Reviewed - Medical Policy Group - Neurology and Neurosurgery. No changes to policy statements. |
| 10/2011 | Reviewed - Medical Policy Group - Gastroenterology, Nutrition and Organ Transplantation. No changes to policy statements. |

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| 11/2011 | Updated to remove statement that ketogenic diets are initiated on an inpatient basis, based on clinical evidence that inpatient initiation does not improve outcomes compared to outpatient initiation. |
| 3/2011 | Reviewed - Medical Policy Group - Allergy and ENT/Otolaryngology. No changes to policy statements. |
| 1/2011 | Reviewed - Medical Policy Group - Neurology and Neurosurgery. No changes to policy statements. |
| 11/2010 | Reviewed - Medical Policy Group - Gastroenterology, Nutrition and Organ Transplantation. No changes to policy statements. |
| 5/2010 | Reviewed - Medical Policy Group - Pediatrics and Endocrinology. No changes to policy statements. |
| 3/2010 | Reviewed - Medical Policy Group - Allergy and ENT/Otolaryngology. No changes to policy statements. |
| 2/2010 | Reviewed - Medical Policy Group - Psychiatry and Ophthalmology. No changes to policy statements. |
| 1/2010 | Reviewed - Medical Policy Group - Neurology and Neurosurgery. No changes to policy statements. |
| 11/2009 | Reviewed - Medical Policy Group - Gastroenterology, Nutrition and Organ Transplantation. No changes to policy statements. |
| 5/2009 | Reviewed - Medical Policy Group - Pediatrics and Endocrinology. No changes to policy statements. |
| 5/2009 | Updated to include the narrative of all enteral formula codes and update of diagnoses: food protein enterocolitis and allergic eosinophilic esophagitis as requested by local experts at the MPG meeting. |
| 3/2009 | Reviewed - Medical Policy Group - Allergy and ENT/Otolaryngology. No changes to policy statements. |
| 2/2009 | Reviewed - Medical Policy Group - Psychiatry and Ophthalmology. No changes to policy statements. |
| 1/2009 | Reviewed - Medical Policy Group - Neurology and Neurosurgery. No changes to policy statements. |
| 11/2008 | Reviewed - Medical Policy Group - Gastroenterology, Nutrition and Organ Transplantation. No changes to policy statements. |
| 8/2008 | Based on the decision from the Implementation sub-BAC committee meeting (August 20, 2008) addressing the recent State mandate change pertaining to a dollar maximum per year for low protein foods the document was edited to remove the reference to a dollar amount allowed for low protein foods. The sub-BAC committee clarified that members should refer to their <i>Benefit Design</i> language to determine benefit coverage rather than this evidenced based medical policy. |
| 5/2008 | Reviewed - Medical Policy Group - Pediatrics and Endocrinology. No changes to policy statements. |
| 3/2008 | Reviewed - Medical Policy Group - Allergy and ENT/Otolaryngology. No changes to policy statements. |
| 2/2008 | Reviewed - Medical Policy Group - Psychiatry and Ophthalmology. No changes to policy statements. |
| 1/2008 | Reviewed - Medical Policy Group - Neurology and Neurosurgery. No changes to policy statements. |
| 11/2007 | Reviewed - Medical Policy Group - Gastroenterology, Nutrition and Organ Transplantation. No changes to policy statements. |
| 5/2007 | Reviewed - Medical Policy Group - Pediatrics and Endocrinology. No changes to policy statements. |
| 3/2007 | Reviewed - Medical Policy Group - Allergy and ENT/Otolaryngology. No changes to policy statements. |
| 2/2007 | Reviewed - Medical Policy Group - Psychiatry and Ophthalmology. No changes to policy statements. |

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| 1/2007 | Reviewed - Medical Policy Group - Neurology and Neurosurgery. No changes to policy statements. |
| 1/2007 | Updated to include coverage for infants under 1 year of age who are intolerant of breast milk or baby formula. |
| 4/1996 | New policy, issued 4/1996, describing covered and non-covered indications. |

References

1. TEC Assessment, 1998; Tab 20
2. Vaisleib II, Buchhalter JR, Zupanc ML. Ketogenic diet: Outpatient initiation, without fluid, or caloric restrictions. *Pediatr Neurol* 2004; 31:198-2002
3. Kwiterovich PO, Vning EP, Pyzik P. Effect of a high-fat ketogenic diet on plasma levels of lipids, lipoproteins, and apolipoproteins in children. *JAMA* 2003; 290:912-20.
4. Liu YM, Williams S, Basualdo-Hammond C et al. A prospective study; growth and nutritional status of children treated with the ketogenic diet. *J Am Diet Assoc* 2003;103:707-12.
5. Vining EP, Pyzik P, McGrogan J et al. Growth of children on the ketogenic diet. *Dev Med Child Neurol* 2002; 44:796-802.
6. Kossoff EH, Krauss GL, McGrogan JR, Freeman JM. Efficacy of the Atkins diet as therapy for intractable epilepsy. *Neurology* 2003; 61:1789-91.

Endnotes

1. Chapter 655 of the Acts of 1983, Massachusetts General Laws, HMO Blue with prescription coverage BCBSMA subscriber certificate. The Mandates states that: "...Coverage shall include those **special formulas** which are approved by the commissioner of the Department of Public Health, prescribed by a physician, and are medically necessary for the treatment of phenylketonuria, tyrosinemia, homocystinuria, maple syrup urine disease, propionic acidemia, or methylmalonic acidemia in infants and children or medically necessary to protect the unborn fetuses of pregnant women with phenylketonuria..."
2. Chapter 683 of the Acts of 1987, Massachusetts General Laws
3. Guidelines submitted and approved 3/94 based the following literature:
 - Recommendations on the Dietary Management of PKU, *Arch Dis Child*; 68:426-7 1993 (Report of the Medical Research Council Working Party on PKU)
 - Effect of Age at Loss of Dietary Control on Intellectual Performance and Behavior of Children with PKU *NEJM*; Vol 314, No. 10, 1986
 - Neurological Deterioration in Young Adults with PKU *Lancet*; 336:602-5.
4. Based upon a 10/96 National Blue Cross Blue Shield Association policy.
5. See the American Academy of Pediatrics Committee on Nutrition recommendations *on Reimbursement for Medical Foods for Inborn Errors of Metabolism*, published in *Pediatrics* vol. 93 No. 5 May 1994.
6. See the American Academy of Pediatrics Committee on Nutrition recommendations *on Soy-Protein Formulas: Recommendations for Use in Infant Feeding*, published in *Pediatrics*, vol. 72 No. 3 September 1983.
7. See the American Academy of Pediatrics Committee on Nutrition recommendations *on Hypoallergenic Infant Formulas*, published in *Pediatrics*, vol. 83 No. 6, June 1989.
8. Adapted from *An Algorithm for Pediatric Enteral Alimentation* by Wilson SE, Dietz, WH, Grand, RJ, *Pediatr Ann* 1987 16:233, thanks to Boston's Childrens Hospital, 1996. Updated 12/00- thanks to Mark Ogino, MD & Jamie Sheldon RD, Massachusetts General Hospital, Boston.
9. See the *Pediatric Nutrition Handbook, Fourth Edition*, by the American Academy of Pediatrics, Ronald Kleinman, MD, Editor.
10. Based on the 10/98 TEC (Technology Evaluation Center) assessment of ketogenic diets for children with refractory epilepsy. This analysis evaluated medical literature through 9/98 to examine the question of whether a ketogenic diet results in a clinically significant reduction in seizure frequency for children who were refractory to anti-epileptic drugs (AEDs). Evidence was in the form of uncontrolled, largely retrospective studies. The definition of "refractory to AEDs" varied between studies; however, in general patients had failed or were intolerant to multiple drug regimens. Subjects in some studies

had a high mean frequency of seizures (for example, 7-13 per day) and a history of extensive prior trials of medications (up to 6-7 different drugs, in some studies). Most studies required an inpatient admission to initiate the diet, to induce a fast, ensure adequate hydration, nutrition, and to educate the patient and family.

11. Based on recommendations from Ronald Kleinman, MD, Massachusetts General Hospital.
12. Based on recommendations from Mark Korson, MD New England Medical Center.
13. Based on recommendations from Eugenia Marcus, MD, President of the Massachusetts Chapter of the American Academy of Pediatrics, MPG 11/00.
14. Adapted from the National Institute of Health Consensus Statement: *PKU Screening and Management*. Volume 17, number 3, October 16-18, 2000.
15. Recommendations on Maternal Phenylketonuria from the American Academy of Pediatrics, 2001:
 - All girls and women of childbearing age with elevated Phe levels, including those with PKU should be identified and counseled regarding risks of maternal PKU fetal effects with uncontrolled blood Phe levels during pregnancy.
 - Women with hyperphenylalaninemia who are unable or unwilling to maintain blood Phe levels should be assisted to obtain adequate means for birth control, including tubal ligation if requested.
 - Women with hyperphenylalaninemia who conceive with blood Phe levels greater than 4-6mg/dL should be counseled regarding the risks to the fetus and offered ultrasonography to detect fetal abnormalities.
 - Women who give birth to infants with features of maternal PKU fetal effects without a known cause should have blood testing for hyperphenylalaninemia.
16. Pediatrics volume 108, number 4 October 2001. *The ketogenic Diet: A 3-to 6-year follow-up of 150 children enrolled prospectively*.
17. New England Journal of Medicine, 1998;338:1715-22. *Long-term prognosis of seizures with onset in childhood*.
18. Recommendations from the MFPC, based on Clinical Pharmacy individual consideration guidelines; January 2004.
19. Ketogenic Diet as a Treatment of Refractory Epilepsy: Based upon the 2004 Blue Cross Blue Shield Association's national policy 2.01.32.
20. Recommendations from local physician experts, March 2009 Medical Policy Group meeting.

To request prior authorization using the Massachusetts Standard Form for Medication Prior Authorization Requests (eForm), click the link below:

<http://www.bluecrossma.org/medical-policies/sites/g/files/csphws2091/files/acquiadam-assets/023%20E%20Form%20medication%20prior%20auth%20instruction%20pm.pdf>