



An Independent Licensee of the Blue Cross Blue Shield Association

EVIDENCE-BASED CRITERIA  
SECTION: MEDICINE

ORIGINAL EFFECTIVE DATE: 09/03/24  
LAST REVIEW DATE: 09/03/24  
CURRENT EFFECTIVE DATE: 09/03/24  
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NEXT ANNUAL REVIEW DATE: 3RD QTR 2025

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## MEDICAL FOODS

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Non-Discrimination Statement and Multi-Language Interpreter Services information are located at the end of this document.

Coverage for services, procedures, medical devices and drugs are dependent upon benefit eligibility as outlined in the member's specific benefit plan. This Evidence-Based Criteria must be read in its entirety to determine coverage eligibility, if any.

This Evidence-Based Criteria provides information related to coverage determinations only and does not imply that a service or treatment is clinically appropriate or inappropriate. The provider and the member are responsible for all decisions regarding the appropriateness of care. Providers should provide BCBSAZ complete medical rationale when requesting any exceptions to these guidelines.

The section identified as "Description" defines or describes a service, procedure, medical device or drug and is in no way intended as a statement of medical necessity and/or coverage.

The section identified as "Criteria" defines criteria to determine whether a service, procedure, medical device or drug is considered medically necessary or experimental or investigational.

State or federal mandates, e.g., FEP program, may dictate that any drug, device or biological product approved by the U.S. Food and Drug Administration (FDA) may not be considered experimental or investigational and thus the drug, device or biological product may be assessed only on the basis of medical necessity.

Evidence-Based Criteria are subject to change as new information becomes available.

For purposes of this Evidence-Based Criteria, the terms "experimental" and "investigational" are considered to be interchangeable.

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## MEDICAL FOODS

### Description:

Medical foods are defined as:

- Modified low protein foods or metabolic formulas, generally available only under the supervision and direction of a physician or a registered nurse practitioner, that are medically necessary for the treatment of an inherited metabolic disorder (also referred to as an inborn error of metabolism) of amino acid, carbohydrate or fat metabolism included in the newborn screening program as required by law, that must be consumed throughout life, and without which the individual may suffer serious mental or physical impairment, or
- An amino acid-based formula that is ordered by a physician or a registered nurse practitioner for an individual diagnosed with an eosinophilic gastrointestinal disorder (EGID)

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### Criteria:

- Medical foods for the treatment of inherited metabolic disorders are considered **medically necessary** based on the benefit plan allowance with documentation of **ALL** of the following:
  1. Diagnosis of **ANY** of the following inherited metabolic disorders included in the newborn screening program:
    - 2,4 Dienoyl-CoA reductase deficiency
    - 2-Methyl-3-hydroxybutyric aciduria or HSD10 disease
    - 3-Hydroxy-3-methylglutaric aciduria
    - 3-Methylcrotonyl-CoA carboxylase deficiency
    - 3-Methylglutaconic aciduria
    - Argininemia
    - Argininosuccinic acidemia
    - Beta-ketothiolase deficiency
    - Biopterin defect in cofactor biosynthesis
    - Biopterin defect in cofactor regeneration
    - Biotinidase deficiency
    - Carnitine-acylcarnitine translocase deficiency
    - Carnitine palmitoyltransferase I deficiency
    - Carnitine palmitoyltransferase II deficiency
    - Carnitine uptake defect
    - Citrullinemia type I
    - Citrullinemia type II
    - Classic galactosemia
    - Galactosepimerase deficiency
    - Galactokinase deficiency
    - Glutaric acidemia type I
    - Glutaric acidemia type II
    - Glycogen storage disease type II or Pompe disease

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- Holocarboxylase synthase deficiency
  - Homocystinuria
  - Hypermethioninemia
  - Hyperphenylalaninemia (benign)
  - Isobutyrylglycinuria
  - Isovaleric acidemia
  - Long-chain 3-hydroxy acyl-CoA dehydrogenase deficiency
  - Malonic acidemia
  - Maple syrup urine disease
  - Medium-chain acyl-CoA dehydrogenase deficiency
  - Medium-chain ketoacyl-CoA thiolase deficiency
  - Medium/short chain L-3 hydroxyacyl-CoA dehydrogenase deficiency
  - Methylmalonic acidemia (cobalamin disorders)
  - Methylmalonic acidemia (mutase deficiency)
  - Methylmalonic acidemia with homocystinuria
  - Mucopolysaccharidosis type I
  - Phenylketonuria
  - Propionic acidemia
  - Trifunctional protein deficiency
  - Tyrosinemia type I
  - Tyrosinemia type II
  - Tyrosinemia type III
  - Very long-chain acyl-CoA dehydrogenase deficiency
  - X-linked adrenoleukodystrophy
2. The medical food is prescribed or ordered and monitored under the supervision of a physician or nurse practitioner
  3. There is risk of mental or physical impairment without the use of the medical food
- Medical foods for the treatment of Eosinophilic Gastrointestinal Disorders (EGID) are considered **medically necessary** based on the benefit plan allowance and documentation of **ALL** of the following:
1. Diagnosis of EGID confirmed by endoscopy **and** biopsy
  2. Individual is under the continuous supervision of a licensed physician or a registered nurse practitioner
  3. The medical food is ordered by a treating provider
  4. There is a risk of mental or physical impairment without the use of the medical food



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- Medical foods for all other indications not previously listed are a **benefit plan exclusion** and **not eligible for coverage**.
- Supplementation of foods, formulas, vitamins or nutrients that are available over the counter (OTC) are a **benefit plan exclusion** and **not eligible for coverage**.

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### Resources:

Literature reviewed 09/03/24. We do not include marketing materials, poster boards and non-published literature in our review.

1. Arizona Administrative Code R9-13-203 Newborn and Infant Bloodspot Tests. Effective September 8, 2022. Accessed July 26, 2024
2. Arizona Revised Statutes, §20-826, §20-826.03, §20-1057, §20-1057.10, §20-2332, §20-2327, §36-694. Accessed July 25, 2024.
3. Baumgartner ER ST. Multiple carboxylase deficiency: inherited and acquired disorders of biotin metabolism. *International Journal for Vitamin and Nutrition Research*.67(5):377-384.
4. Blue Cross Blue Shield of Arizona. Benefit Plan Booklet. January 1, 2024. Accessed July 25, 2024.
5. Defendi GL. Biotinidase Deficiency. Medscape. Updated February 15, 2023. Accessed July 1, 2024. <https://emedicine.medscape.com/article/942055-overview?form=fpf#a4>.
6. Gillingham MB, Connor WE, Matern D, et al. Optimal dietary therapy of long-chain 3-hydroxyacyl-CoA dehydrogenase deficiency. *Mol Genet Metab*. Jun 2003;79(2):114.
7. Health Resources & Services Administration (HRSA). HRSA Newborn Screening. Accessed August 16, 2024. <https://newbornscreening.hrsa.gov/>

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### Coding:

HCPCS: B4153, B4155, B4157, B4161, B4162, B4168, B4172, B4176, B4178, B4189, B4193, B4197, B4199, B9002, B9004, B9006, B9998, B9999, J3490, S9433, S9434, S9435

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## **MEDICAL FOODS**

| <b><u>History:</u></b>         | <b><u>Date:</u></b> | <b><u>Activity:</u></b> |
|--------------------------------|---------------------|-------------------------|
| Medical Policy Panel           | 09/03/24            | Approved guideline      |
| Medical Director (Dr. Sutanto) | 08/22/24            | Review with revisions   |
| Legal Department               | 08/15/24            | Review with revisions   |
| Medical Director (Dr. Raja)    | 08/05/24            | Review with revisions   |
| Legal Department               | 07/25/24            | Review with revisions   |

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### **Policy Revisions:**



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### Non-Discrimination Statement:

Blue Cross Blue Shield of Arizona (BCBSAZ) complies with applicable Federal civil rights laws and does not discriminate on the basis of race, color, national origin, age, disability or sex. BCBSAZ provides appropriate free aids and services, such as qualified interpreters and written information in other formats, to people with disabilities to communicate effectively with us. BCBSAZ also provides free language services to people whose primary language is not English, such as qualified interpreters and information written in other languages. If you need these services, call (602) 864-4884 for Spanish and (877) 475-4799 for all other languages and other aids and services.

If you believe that BCBSAZ has failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability or sex, you can file a grievance with: BCBSAZ's Civil Rights Coordinator, Attn: Civil Rights Coordinator, Blue Cross Blue Shield of Arizona, P.O. Box 13466, Phoenix, AZ 85002-3466, (602) 864-2288, TTY/TDD (602) 864-4823, [crc@azblue.com](mailto:crc@azblue.com). You can file a grievance in person or by mail or email. If you need help filing a grievance BCBSAZ's Civil Rights Coordinator is available to help you. You can also file a civil rights complaint with the U.S. Department of Health and Human Services, Office for Civil Rights electronically through the Office for Civil Rights Complaint Portal, available at <https://ocrportal.hhs.gov/ocr/portal/lobby.jsf>, or by mail or phone at: U.S. Department of Health and Human Services, 200 Independence Avenue SW., Room 509F, HHH Building, Washington, DC 20201, 1-800-368-1019, 800-537-7697 (TDD). Complaint forms are available at <http://www.hhs.gov/ocr/office/file/index.html>

### Multi-Language Interpreter Services:

Spanish: Si usted, o alguien a quien usted está ayudando, tiene preguntas acerca de Blue Cross Blue Shield of Arizona, tiene derecho a obtener ayuda e información en su idioma sin costo alguno. Para hablar con un intérprete, llame al 602-864-4884.

Navajo: Díí kwe'é atah nilínígíí Blue Cross Blue Shield of Arizona haada yit'éego bina'idíłkídkgo éí doodago Háida bíjá anilyeedígíí t'áadoo le'é yina'idíłkídkgo beehaz'áanii hółqo díí t'áa hazaadk'ehjí háká a'doowolgo bee haz'ą doo baqah ilínígóó. Ata' halne'ígíí kojí' bich'í' hodíłnih 877-475-4799.

Chinese: 如果您，或是您正在協助的對象，有關於插入項目的名稱 Blue Cross Blue Shield of Arizona 方面的問題，您有權利免費以您的母語得到幫助和訊息。洽詢一位翻譯員，請撥電話 在此插入數字 877-475-4799。

Vietnamese: Nếu quý vị, hay người mà quý vị đang giúp đỡ, có câu hỏi về Blue Cross Blue Shield of Arizona quý vị sẽ có quyền được giúp và có thêm thông tin bằng ngôn ngữ của mình miễn phí. Để nói chuyện với một thông dịch viên, xin gọi 877-475-4799.

Arabic:

إن كان لديك أو لدى شخص تساعد أسئلة بخصوص Blue Cross Blue Shield of Arizona، فلديك الحق في الحصول على المساعدة والمعلومات الضرورية بلغتك من دون أية تكلفة. للتحدث مع مترجم اتصل بـ 877-475-4799.



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Multi-Language Interpreter Services:

Tagalog: Kung ikaw, o ang iyong tinutulangan, ay may mga katanungan tungkol sa Blue Cross Blue Shield of Arizona, may karapatan ka na makakuha ng tulong at impormasyon sa iyong wika ng walang gastos. Upang makausap ang isang tagasalin, tumawag sa 877-475-4799.

Korean: 만약 귀하 또는 귀하가 돕고 있는 어떤 사람이 Blue Cross Blue Shield of Arizona 에 관해서 질문이 있다면 귀하는 그러한 도움과 정보를 귀하의 언어로 비용 부담없이 얻을 수 있는 권리가 있습니다. 그렇게 통역사와 얘기하기 위해서는 877-475-4799 로 전화하십시오.

French: Si vous, ou quelqu'un que vous êtes en train d'aider, a des questions à propos de Blue Cross Blue Shield of Arizona, vous avez le droit d'obtenir de l'aide et l'information dans votre langue à aucun coût. Pour parler à un interprète, appelez 877-475-4799.

German: Falls Sie oder jemand, dem Sie helfen, Fragen zum Blue Cross Blue Shield of Arizona haben, haben Sie das Recht, kostenlose Hilfe und Informationen in Ihrer Sprache zu erhalten. Um mit einem Dolmetscher zu sprechen, rufen Sie bitte die Nummer 877-475-4799 an.

Russian: Если у вас или лица, которому вы помогаете, имеются вопросы по поводу Blue Cross Blue Shield of Arizona, то вы имеете право на бесплатное получение помощи и информации на вашем языке. Для разговора с переводчиком позвоните по телефону 877-475-4799.

Japanese: ご本人様、またはお客様の身の回りの方でも、Blue Cross Blue Shield of Arizona についてご質問がございましたら、ご希望の言語でサポートを受けたり、情報を入手したりすることができます。料金はかかりません。通訳とお話される場合、877-475-4799 までお電話ください。

Farsi:

اگر شما، یا کسی که شما به او کمک میکنید، سوال در مورد Blue Cross Blue Shield of Arizona، داشته باشید حق این را دارید که کمک و اطلاعات به زبان خود را به طور رایگان دریافت نمایید 877-475-4799 [تماس حاصل نمایید.]

Assyrian:

Blue Cross Blue Shield of Arizona... 877-475-4799

Serbo-Croatian: Ukoliko Vi ili neko kome Vi pomažete ima pitanje o Blue Cross Blue Shield of Arizona, imate pravo da besplatno dobijete pomoć i informacije na Vašem jeziku. Da biste razgovarali sa prevodiocem, nazovite 877-475-4799.

Thai: หากคุณ หรือคนที่คุณกำลังช่วยเหลือถามเกี่ยวกับ Blue Cross Blue Shield of Arizona คุณมีสิทธิที่จะได้รับความช่วยเหลือและข้อมูลในภาษา ของคุณได้โดยไม่มีค่าใช้จ่าย โปรดขอยืม โทร 877-475-4799