PART B PRIOR AUTHORIZATION CRITERIA FOR APPROVAL

Initial Evaluation:
Givlaari (givosiran) will be approved when ALL of the following are met:

1. The patient is 18 years of age or older;
   AND
2. The patient has a confirmed diagnosis of **acute hepatic porphyria (AHP)** (including acute intermittent porphyria, hereditary coproporphyria, variegate porphyria, aminolevulinic acid (ALA) dehydratase deficient porphyria) **[medical record documentation required]**;
   AND
3. The patient has documentation of elevated urinary or plasma porphobilinogen (PBG) or ALA values **[medical record documentation required]**;
4. **AND**
5. The patient has had one of the following **[medical record documentation required]**:
   a. History of at least two documented porphyria attacks within the 6 months prior to initiation of therapy with the requested agent (requiring hospitalization, urgent healthcare visit, or intravenous hemin administration at home);
      OR
   b. History of one severe attack within the past year with central nervous system (CNS), autonomic nervous system (ANS), or peripheral nervous system (PNS) involvement (e.g., hallucinations, seizures, respiratory failure, paralysis);
   c. **AND**
6. The patient has not had and is not anticipating a liver transplantation **[medical record documentation required]**;
   AND
7. The prescriber is a specialist in the area of the patient's diagnosis (e.g., hepatologist, hematologist, neurologist) or has consulted with a specialist in the area of the patient's diagnosis **[medical record documentation required].**

Duration of Approval: 12 months

Renewal Evaluation:
Givlaari (givosiran) will be approved when ALL of the following are met:

1. The patient would have met initial criteria for approval at the time they started therapy **[medical record documentation required]**;
   AND
2. The patient has had a positive clinical response while using the requested agent, as demonstrated by a reduction in porphyria attacks requiring hospitalization, urgent healthcare visit, or intravenous hemin administration **[medical record documentation required]**;
   AND

3. The prescriber is a specialist in the area of the patient's diagnosis (e.g., hepatologist, hematologist, neurologist) or has consulted with a specialist in the area of the patient's diagnosis **[medical record documentation required].**

Duration of Approval: 12 months

FDA Label Reference			
Medication	**Indication**	**Dosing**	**HCPCS**
givosiran (Givlaari®) subcutaneous (SC) injection	Acute hepatic porphyria (AHP) in adults	SC: 2.5 mg/kg once monthly	J0223

References: all information referenced is from FDA package insert unless otherwise noted below.

1. Stölzel U, Doss MO, Schuppan D, et al. Clinical guide and update on porphyrias. *Gastroenterology*. 2019;157(2):365-81.
2. Kauppinen R. Porphyrias. *Lancet*. 2005;365:241-52.

Policy Implementation/Update Information:

- **August 2025:** 8/1/2025 policy implementation

Non-Discrimination and Accessibility Notice

Discrimination is Against the Law

- Blue Cross and Blue Shield of North Carolina ("Blue Cross NC") complies with applicable Federal civil rights laws and does not discriminate on the basis of race, color, national origin, age, disability, or sex.
- Blue Cross NC does not exclude people or treat them differently because of race, color, national origin, age, disability, or sex.

  Blue Cross NC:

  - Provides free aids and services to people with disabilities to communicate effectively with us, such as:
    - Qualified interpreters
    - Written information in other formats (large print, audio, accessible electronic formats, other formats)
  - Provides free language services to people whose primary language is not English, such as:
    - Qualified interpreters
    - Information written in other languages

- If you need these services, contact Customer Service **1-888-206-4697**, TTY and TDD, call **1-800-442-7028**.

- If you believe that Blue Cross NC 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:
  - Blue Cross NC, PO Box 2291, Durham, NC 27702, Attention: Civil Rights Coordinator- Privacy, Ethics & Corporate Policy Office, Telephone **919-765-1663**, Fax **919-287-5613**, TTY **1-888-291-1783** civilrightscoordinator@bcbsnc.com

- You can file a grievance in person or by mail, fax, or email. If you need help filing a grievance, Civil Rights Coordinator - Privacy, Ethics & Corporate Policy Office 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, D.C. 20201 **1-800-368-1019, 800-537-7697** (TDD). Complaint forms are available at *http://www.hhs.gov/ocr/office/file/index.html*.

- This Notice and/or attachments may have important information about your application or coverage through Blue Cross NC. Look for key dates. You may need to take action by certain deadlines to keep your health coverage or help with costs. You have the right to get this information and help in your language at no cost. Call Customer Service **1-888-206-4697**.

ATTENTION: If you speak another language, language assistance services, free of charge, are available to you. Call 1-888-206-4697 (TTY: 1-800-442-7028).

ATENCIÓN: Si habla español, tiene a su disposición servicios gratuitos de asistencia lingüística. Llame al 1-888-206-4697 (TTY: 1-800-442-7028).

□ □： 如果您講廣東話或普通話□ □ □ □ □ □ □ □ □ □ □ □ □ □ □ □ □ □ 1-888-206-4697 (TTY：1-800-442-7028）。

CHÚ Ý: Nếu bạn nói Tiếng Việt, có các dịch vụ hỗ trợ ngôn ngữ miễn phí dành cho bạn. Gọi số 1-888-206-4697 (TTY: 1-800-442-7028).

□ □:□ □ □ □ □ □ □ □ □ □ □,□ □. 1-888-206-4697 (TTY: 1- 800-442-7028)□ □ □ □ □ □ □ □ □ □.

ATTENTION : Si vous parlez français, des services d'aide linguistique vous sont proposés gratuitement. Appelez le 1-888-206-4697 (ATS : 1-800-442-7028).

ملحوظة: إذا كنت تتحدث اللغة العربية، فإن خدمات المساعدة اللغوية تتوافر لك بالمجان. اتصل برقم 1-888-206-4697. المبرقة الكاتبة: 1-800-442-7028.

LUS CEEV: Yog tias koj hais lus Hmoob, cov kev pab txog lus, muaj kev pab dawb rau koj. Hu rau 1-888-206-4697 (TTY: 1-800-442-7028).

ВНИМАНИЕ: Если вы говорите на русском языке, то вам доступны бесплатные услуги перевода. Звоните 1-888-206-4697 (телетайп: 1-800-442-7028).

PAUNAWA: Kung nagsasalita ka ng Tagalog, maaari kang gumamit ng mga serbisyo ng tulong sa wika nang walang bayad. Tumawag sa 1-888-206-4697 (TTY: 1-800-442-7028).

સુચના: જો તમે ગુજરાતી બોલતા હો, તો નિ:સુલ્કુ ભાષા સહાય સેવાઓ તમારા માટે ઉપલબ્ધ છે. ફોન કરો 1-888-206-4697 (TTY: 1-800-442-7028).

1-888-206-4697 (TTY: 1-800-442-7028)Б

ACHTUNG: Wenn Sie Deutsch sprechen, stehen Ihnen kostenlos sprachliche Hilfsdienstleistungen zur Verfügung. Rufnummer: 1-888-206-4697 (TTY: 1-800-442-7028).

1-888-206-4697 (TTY: 1-800-442-7028)

1-888-206-4697 (TTY: 1-800-442-7028).

注意事項：日本語を話される場合、無料の言語支援をご利用いただけます。1-888-206-4697（TTY: 1-800-442-7028）まで、お電話にてご連絡ください。