Maryland
DEPARTMENT OF HEALTH

Givlaar (givosiarn)

Givlaar is an aminolevulinate synthase 1-directed small interfering RNA indicated for the treatment of adults with acute hepatic porphyria (AHP).

I. Criteria for Initial Approval

Givlaar will be considered for coverage when **ALL** of the criteria below are met, confirmed with supporting medical documentation.

- Patient is ≥ 18 years of age;
- Patient has a confirmed diagnosis of AHP (including acute intermittent porphyria, hereditary coproporphyria, variegate porphyria, aminolevulinic acid (ALA) dehydratase deficient porphyria);
- Documentation of elevated urinary or plasma porphobilinogen (PBG) or ALA values;
- Patient has had one of the following:
    - History of at least two documented porphyria attacks within the 6 months prior to initiation (requiring hospitalization, urgent healthcare visit, or intravenous hemin administration at home); OR
    - 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);
- Givosiran is prescribed by or in consultation with a specialist in porphyria treatment (e.g., hepatologist, hematologist, neurologist);
- Patient has not had, nor is anticipating, a liver transplantation;
- The patient has been advised to avoid the following medications and is not currently prescribed any CYP1A2 and CYP2D6 substrates such as:
    - Alosetron, caffeine, duloxetine, melatonin, ramelteon, tasimelteon, tizanidine clozapine, pirfenidone, ramosetron, theophylline; or
    - Atomoxetine, desipramine, dextromethorphan, eliglustat, nebivolol, nortriptyline, perphenazine, tolterodine, R-venlafaxine encainide, imipramine, metoprolol, propafenone, propranolol, tramadol, trimipramine, S-venlafaxine; and

- Provider agrees to screen and monitor liver and renal function during the course of therapy.

II. Criteria for Continuation of Therapy

All of the criteria for initial therapy (in **Section I.**) must be met **AND** the provider attests to a positive clinical response.

- Documentation that the patient has experienced a positive clinical response by demonstrating the following from pretreatment baseline:
    - Reduction in hemin administration requirements (if previously required, including prophylactic and/or treatment doses);
    - Reduction in the rate and/or number of porphyria attacks;
    - Improvement of signs and symptoms of AHPs (e.g., pain, neurological, gastrointestinal, renal, quality of life, etc.); and
- Provider attests that the:
    - Patient will not receive concomitant prophylactic hemin treatment while on Givlaari; and
    - Patient has not had a liver transplant.

III. Dosing/Administration

Givlaar must be administered according to the current FDA labeling guidelines for dosage and timing. The recommended dosing is as follows:

- The recommended dose of Givlaari is 2.5 mg/kg once monthly by subcutaneous injection.

IV. Length of Authorization For initial therapy

Givlaar will be authorized for 6 months when criteria for initial approval are met. Continuing therapy with Givlaar will be authorized for 12 months.

V. Billing Code/Information

J0223 – Injection, givosiran, 0.5 mg: 1 billable unit = 0.5 mg

Prior authorization of benefits is not the practice of medicine nor the substitute for the independent medical judgment of a treating medical provider. The materials provided are a component used to assist in making coverage decisions and administering benefits. Prior authorization does not constitute a contract or guarantee regarding member eligibility or payment. Prior authorization criteria are established based on a collaborative effort using input from the current medical literature and based on evidence available at the time.

Approved by MDH Clinical Criteria Committee: 9/28/2021
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