BlueCross BlueShield Arizona

An Independent Licensee of the Blue Cross Blue Shield Association

PHARMACY COVERAGE GUIDELINE

DIACOMIT® (stiripentol)
Generic Equivalent (if available)

This Pharmacy Coverage Guideline (PCG):
- Provides information about the reasons, basis, and information sources we use for coverage decisions
- Is not an opinion that a drug (collectively "Service") is clinically appropriate or inappropriate for a patient
- Is not a substitute for a provider's judgment (Provider and patient are responsible for all decisions about appropriateness of care)
- Is subject to all provisions e.g. (benefit coverage, limits, and exclusions) in the member's benefit plan; and
- Is subject to change as new information becomes available.

Scope
- This PCG applies to Commercial and/or Marketplace plans
- This PCG does not apply to the Federal Employee Program, Medicare Advantage, Medicaid or members of out-of-state Blue Cross and/or Blue Shield Plans

Instructions & Guidance
- To determine whether a member is eligible for the Service, read the entire PCG.
- This PCG is used for FDA approved indications including, but not limited to, a diagnosis and/or treatment with dosing, frequency, and duration.
- Use of a drug outside the FDA approved guidelines, refer to the appropriate Off-Label Use policy.
- The "Criteria" section outlines the factors and information we use to decide if the Service is medically necessary as defined in the Member's benefit plan.
- The "Description" section describes the Service.
- The "Definition" section defines certain words, terms or items within the policy and may include tables and charts.
- The "Resources" section lists the information and materials we considered in developing this PCG
- **We do not accept patient use of samples as evidence of an initial course of treatment, justification for continuation of therapy, or evidence of adequate trial and failure.**
- Information about medications that require prior authorization is available at www.azblue.com/pharmacy. You must fully complete the request form and provide chart notes, lab workup and any other supporting documentation. The prescribing provider must sign the form. Fax the form to BCBSAZ Pharmacy Management at (602) 864-3126 or email it to Pharmacyprecert@azblue.com.

Medical Necessity Requirements for DIACOMIT (stiripentol)

Criteria for Initial Therapy:

Prescriber Qualifications
- Prescribed by a physician specializing in the diagnosis or in consultation with a Neurologist

Indication
- Diagnosis of seizures associated with Dravet syndrome
- Experiencing at least four generalized clonic or tonic clonic seizures per month despite treatment with clobazam and valproate

ORIGINAL EFFECTIVE DATE: 05/19/2019 | ARCHIVE DATE: | LAST REVIEW DATE: 05/21/2026 | LAST CRITERIA REVISION DATE: 05/16/2024

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Age Requirement
- 6 months of age or older and weighs at least 7 kilograms

Baseline Clinical Evaluation
- Completed baseline complete blood count test before starting treatment
- Will not be used as monotherapy; will be used with clobazam and valproate

Brand Specific Criteria
- Have failure, contraindication, or intolerance with **THREE** generic equivalents (if available) for at least three months each. **Note:** Any failure, contraindication, or intolerance to the generic drugs should be reported to the FDA (see Definitions section)

Safety
- Does not have moderate or severe renal impairment
- Does not have moderate or severe hepatic impairment

Documentation Requirements
- A completed request form must be submitted including:
  - Chart notes
  - Lab results (CBC)
  - Supporting clinical documentation

Initial Therapy Criteria Approval Duration
- 6 months OR end of plan year

Criteria for Continuation of Therapy (renewal therapy):
Note: Manufacturer assistance (e.g., coupons, samples, etc.) are not considered for continuation of therapy

Prescriber Qualification
- Continues to be seen by a physician specializing in or in consultation with a Neurologist

Clinical Response
- No evidence of disease progression
- Achieved and maintains at least a 50 percent decrease in frequency (per 30 days) of generalized clonic or tonic clonic seizures compared to baseline
- Achieved and maintains a longer interval between seizures over baseline

Adherence
- Adherence to the prescribed therapy regimen has been documented

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Brand Specific Criteria

- Have failure, contraindication, or intolerance with **THREE** generic equivalents (if available) for at least three months each. **Note:** Any failure, contraindication, or intolerance to the generic drugs should be reported to the FDA (see Definitions section)

Safety

- No significant adverse drug effects such as:
  - Emergence or worsening of depression, suicidal thoughts, behavior, or thoughts of self harm
  - Any unusual changes in mood or behavior
  - Phenylketonuria from use of powder for suspension formulation
  - Neutropenia
  - Thrombocytopenia

Documentation Requirements

- Chart notes
- Supporting clinical documentation with evidence of improvement in given indication
- Lab values that confirm safe use

Continuation Therapy Criteria Approval Duration

- 12 months OR end of plan year

Criteria for Off-Label Use Requests:

Criteria for a request for non-FDA use or indication, treatment with dosing, frequency, or duration outside the FDA-approved dosing, frequency, and duration, refer to one of the following Pharmacy Coverage Guideline:

1. Off-Label Use of Non-Cancer Medications

2. Off-Label Use of Cancer Medications

Description:

Diacomit (stiripentol) is indicated for the treatment of seizures associated with Dravet syndrome (DS) in patients taking clobazam who are 6 months of age and older and weighing 7 kilograms or more. There are no clinical data to support the use of Diacomit (stiripentol) as monotherapy in DS. The mechanism by which Diacomit (stiripentol) exerts its anticonvulsant effect in humans is unknown. Possible mechanisms of action include direct effects mediated through the gamma-aminobutyric acid (GABA) receptor and indirect effects involving inhibition of cytochrome P450 activity with resulting increase in blood levels of clobazam and its active metabolite.

DS, previously known as severe myoclonic epilepsy of infancy, is a rare early-onset epileptic encephalopathy characterized by refractory epilepsy and neurodevelopmental problems beginning in infancy. Patients present in the first year of life with a prolonged, often febrile, clonic seizure in the setting of normal cognitive and motor

development prior to seizure onset. In most, febrile, and afebrile seizures, including episodes of status epilepticus, recur repeatedly in the weeks to months after the initial event, and psychomotor impairment begins thereafter. Myoclonus, both epileptic and non-epileptic, occurs frequently. The majority of older children and young adults with DS have motor system dysfunction, gait and postural abnormalities, and cognitive and behavioral impairment.

DS seizures tend to be refractory to most anti-seizure drugs, and some patients derive benefit from a ketogenic diet and vagus nerve stimulation. The most commonly used anti-seizure drugs include valproate, clobazam, topiramate, levetiracetam, stiripentol, and cannabidiol. Most patients require two or more agents to achieve reasonable seizure control.

A recent international consensus panel in 2022 ranked maintenance therapy as follows: first line: valproate; second line: clobazam, fenfluramine, and stiripentol; third line: cannabidiol; fourth line: topiramate, ketogenic diet. However, the separation of the categories was not universally agreed upon. Some argued that clobazam, fenfluramine, and stiripentol should be first line. Others argued that cannabidiol and fenfluramine should be considered at any point. It was agreed upon that carbamazepine, oxcarbazepine, lamotrigine, and phenytoin should be avoided.

Definitions:

U.S. Food and Drug Administration (FDA) MedWatch Forms for FDA Safety Reporting
MedWatch Forms for FDA Safety Reporting | FDA

Resources:

Diacomit (stiripentol) cap & powder for oral solution product information, revised by Biocodex, Inc. 06-2024. Available at DailyMed http://dailymed.nlm.nih.gov. Accessed on February 12, 2026.

Andrade DM, Nascimento FA. Dravet syndrome: Genetics, clinical features, and diagnosis. In: UpToDate, Nordil DR, Dashe JF (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at http://uptodate.com. Literature current through March 2026. Topic last updated October 03, 2025. Accessed on April 27, 2026.

Andrade DM, Nascimento FA. Dravet syndrome: Management and prognosis. In: UpToDate, Nordil DR, Dashe JF (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at http://uptodate.com. Literature current through March 2026. Topic last updated April 08, 2026. Accessed on April 27, 2026.

Wirrell EC, Hood V, Knupp KG, et al.: International consensus on diagnosis and management of Dravet syndrome. Epilepsia 2022;63:1761-1777. Available at https://doi.org/10.1111/epi.17274. Accessed on March 16, 2023. Re-evaluated April 27, 2026.