# Drug Coverage Policy

**Effective Date.....5/15/2025**
**Coverage Policy Number..... IP0409**
**Policy Title.....Diacomit**

# Antiseizure Medications – Diacomit

- Diacomit® (stiripentol capsules and powder for oral suspension – Biocodex)

***INSTRUCTIONS FOR USE***
*The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment where appropriate and have discretion in making individual coverage determinations. Where coverage for care or services does not depend on specific circumstances, reimbursement will only be provided if a requested service(s) is submitted in accordance with the relevant criteria outlined in the applicable Coverage Policy, including covered diagnosis and/or procedure code(s). Reimbursement is not allowed for services when billed for conditions or diagnoses that are not covered under this Coverage Policy (see "Coding Information" below). When billing, providers must use the most appropriate codes as of the effective date of the submission. Claims submitted for services that are not accompanied by covered code(s) under the applicable Coverage Policy will be denied as not covered. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.*

**OVERVIEW**
Diacomit, an antiseizure medication (ASM), is indicated for the treatment of seizures associated with **Dravet syndrome** in patients ≥ 6 months of age and weighing ≥ 7 kg taking clobazam.[1] There are no clinical data to support the use of Diacomit as monotherapy in Dravet syndrome.

**Disease Overview**
Dravet syndrome is a rare genetic epileptic encephalopathy (dysfunction of the brain) marked with frequent and/or prolonged seizures.[2,3] The seizures generally begin in the first year of life in an otherwise healthy infant. Affected individuals can develop many seizure types: myoclonic, tonic-clonic, absence, atypical absence, atonic, focal aware or impaired awareness (previously called partial seizures), and status epilepticus.[3] Two or more ASMs are often needed to control the seizures; most of the seizures are refractory to medications. The goals of treatment are cessation of prolonged convulsions, reduction in overall seizure frequency, and minimization of treatment side effects.[4,5]

**Clinical Efficacy in Other Refractory Seizures**
In one study (n = 212), Diacomit was studied in children with different types of epilepsy syndromes (including Lennox-Gastaut Syndrome [LGS]; infantile spasms; infection-related or anoxo-ischemic epilepsy syndromes; tuberous sclerosis complex; Sturge-Weber syndrome; Doose syndrome; cortical malformation/dysplasia; and epilepsy with myoclonic absences) whose seizures were refractory to more than two ASMs (including vigabatrin).[6] In the 88 patients who completed the 3-month placebo-controlled study, 56.8% of patients with partial epilepsy responded (with 14% becoming seizure-free) compared with 41.9% of patients with generalized epilepsy and 38.4% of patients with myoclonic epilepsy. Diacomit has also been administered to patients with refractory epileptic encephalopathies due to etiologies other than Dravet syndrome.[7] A single-blind, exploratory trial evaluated Diacomit in combination with standard treatment in 16 patients with LGS and 8 patients with symptomatic generalized epilepsy of the Lennox-Gastaut type.[8] There were 15 evaluable patients with LGS. The overall results identified some benefit for LGS where 60% of patients were responders (based on 50% responder rate). Diacomit treatment produced a mean 62% seizure reduction and a median 80% reduction from baseline. Additionally, a published study of Diacomit added to carbamazepine in childhood partial epilepsy (n = 67) demonstrated seizure response in 32 patients with conditions including herpetic encephalitis, LGS, and tuberous sclerosis complex.[9]

**Guidelines/Recommendations**
At this time, there are three drugs approved for the treatment of seizures associated with Dravet syndrome: Diacomit, Epidiolex® (cannabidiol oral solution), and Fintepla® (fenfluramine oral solution).[1,10,11] An expert panel considers valproic acid and clobazam to be the first-line treatment for Dravet syndrome.[5] If seizure control is suboptimal, Diacomit and topiramate are second-line treatment. Ketogenic diet is moderately effective and can also be considered second-line. The Dravet Foundation states that Diacomit, Epidiolex, and Fintepla are considered first-line agents for the treatment of Dravet syndrome.[2] If control is still inadequate, other therapies to consider are clonazepam, levetiracetam, and zonisamide.[2,4,5] Sodium channel blockers (e.g., carbamazepine, oxcarbazepine, lamotrigine, and phenytoin) can worsen seizures in Dravet syndrome. Additionally, vigabatrin and tiagabine may increase the frequency of myoclonic seizures and should be avoided.

## Coverage Policy

**POLICY STATEMENT**
Prior Authorization is required for prescription benefit coverage of Diacomit. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with Diacomit as well as the monitoring required for adverse events and long-term efficacy, initial approval requires Diacomit to be prescribed by or in consultation with a physician who specializes in the condition being treated.

**Stiripentol (Diacomit) is considered medically necessary when ONE of the following is met:**

**FDA-Approved Indication**

1. **Dravet Syndrome.** Approve for 1 year if the patient meets ONE of the following (A or B):
   - **A)** Initial Therapy: Approve if the patient meets the following (i, ii, and iii):
     - **i.** Patient is ≥ 6 months of age and weighs ≥ 7 kg; AND
     - **ii.** Patient meets ONE of the following (a or b):
       - **a)** Patient is taking concomitant clobazam; OR
       - **b)** Patient is unable to take clobazam due to adverse events as determined by the prescriber; AND
     - **iii.** The medication is prescribed by or in consultation with a neurologist; OR
   - **B)** Patient is Currently Receiving Diacomit: Approve if the patient is responding to therapy (e.g., reduced seizure severity, frequency, and/or duration) as determined by the prescriber.

**Other Uses with Supportive Evidence**

2. **Treatment-Refractory Seizures/Epilepsy (specific rare conditions)** [i.e., Lennox-Gastaut Syndrome; infantile spasms; tuberous sclerosis complex; Sturge-Weber syndrome; Doose syndrome; infection-related or anoxo-ischemic epilepsy syndromes; cortical malformation/dysplasia; epileptic encephalopathies associated with sodium channel mutations; and epilepsy with myoclonic absences]. Approve for 1 year if the patient meets ONE of the following (A or B):
   - **A)** Initial Therapy: Approve if the patient meets the following (i, ii, and iii):
     - **i.** Patient is ≥ 6 months of age and weighs ≥ 7 kg; AND
     - **ii.** Patient has tried at least two other antiseizure medications; AND
       Note: Examples of other antiseizure medications include valproic acid, lamotrigine, topiramate, clonazepam, Banzel® (rufinamide tablet, oral suspension), felbamate, clobazam, Fycompa® (perampanel tablet, oral suspension), vigabatrin, levetiracetam, zonisamide.
     - **iii.** The medication is prescribed by or in consultation with a neurologist; OR
   - **B)** Patient is Currently Receiving Diacomit: Approve if the patient is responding to therapy (e.g., reduced seizure severity, frequency, and/or duration) as determined by the prescriber.

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Receipt of sample product does not satisfy any criteria requirements for coverage.

**Diacomit for any other use is considered not medically necessary. Criteria will be updated as new published data are available.**

## References


1. Diacomit® capsules and oral suspension [prescribing information]. Redwood City, CA: Biocodex; July 2022.
2. Dravet Foundation – Dravet Syndrome. Available at: https://www.dravetfoundation.org/what-is-dravet-syndrome/. Accessed on February 14, 2025.
3. Shafer PO. Epilepsy Foundation – Dravet Syndrome. Updated August 2020. Available at: https://www.epilepsy.com/learn/types-epilepsy-syndromes/dravet-syndrome/. Accessed on February 14, 2025.

4. Knupp KG, Wirrell EC. Treatment Strategies for Dravet Syndrome. *CNS Drugs*. 2018;32(4):335-350.
5. Wirrell EC, Laux L, Donner, et al. Optimizing the diagnosis and management of Dravet syndrome: recommendations from a North American Consensus Panel. *Pediatr Neurol*. 2017;68:18-34.
6. Perez J, Chiron C, Musial C, et al. Stiripentol: efficacy and tolerability in children with epilepsy. *Epilepsia*. 1999;40(11):1618-1626.
7. Gil-Nagel A, Aledo-Serrano A, Beltrán-Corbellini Á, et al. Efficacy and tolerability of add-on stiripentol in real-world clinical practice: An observational study in Dravet syndrome and non-Dravet developmental and epileptic encephalopathies. *Epilepsia Open*. 2024;9(1):164-175.
8. Center for Drug Evaluation and Research. Clinical review of Diacomit. Available at: https://www.accessdata.fda.gov/drugsatfda_docs/nda/2018/206709Orig1s000,207223Orig1s000MedR.pdf. Accessed on February 14, 2025.
9. Chiron C, Tonnelier S, Rey E, et al. Stiripentol in childhood partial epilepsy: randomized placebo-controlled trial with enrichment and withdrawal design. *J Child Neurol*. 2006;21(6):496-502.
10. Epidiolex® oral solution [prescribing information]. Palo Alto, CA: Jazz; March 2024.
11. Fintepla® oral solution [prescribing information]. Smyrna, GA: UCB; December 2023.

## Revision Details

| Type of Revision | Summary of Changes | Date |
|---|---|---|
| Annual Review | No content criteria changes | 5/1/2024 |
| Annual Revision | No criteria changes. | 5/15/2025 |

The policy effective date is in force until updated or retired.