Executive Summary

Purpose: The MO HealthNet Pharmacy Program will implement a state-specific preferred drug list.

Why Issue Selected: As of June 2020, the FDA has granted authorization of three pharmacologic therapies for the treatment of Dravet syndrome: Diacomit®, Epidiolex®, and Fintepla®. Dravet syndrome, a rare genetic form of epilepsy afflicting 1 in 15,700 births in the United States appears during the first year of life in otherwise healthy infants as a prolonged seizure with fever. Quality of life for an individual with Dravet syndrome can be severely impacted as the condition is often associated with a multitude of comorbidities including frequent and prolonged seizures, developmental delays, and chronic infections. Diacomit, which must be administered with clobazam, is approved for use in patients 6 months of age or older while Epidiolex is approved down to 1 year of age and Fintepla down to 2 years of age. Fintepla is only available through a REMS program that requires echocardiograms before, during, and after therapy.

Epidiolex and Fintepla also share the indication of Lennox-Gastaut syndrome (LGS), but Epidiolex remains the only agent in the class indicated for Tuberous sclerosis complex (TSC). While LGS is also a form of childhood-onset epilepsy, TSC is a genetic condition that results in the formation of non-cancerous tumors in various parts of the body such as the brain, kidney, heart, eyes, lungs, and skin. Both are considered rare conditions with LGS accounting for 1-4% of all childhood epilepsy cases and TSC affecting approximately 40,000 to 80,000 individuals in the United States. Although all three agents appear to exert their therapeutic effects via varying methods, their exact mechanisms of actions are not thoroughly understood.

Total program savings for the PDL classes will be regularly reviewed.

Program-Specific Information:

<table>
<thead>
<tr>
<th>Preferred Agents</th>
<th>Non-Preferred Agents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epidiolex®</td>
<td>Diacomit®</td>
</tr>
<tr>
<td></td>
<td>Fintepla®</td>
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</tbody>
</table>

Type of Criteria:
- [ ] Increased risk of ADE
- [x] Preferred Drug List
- [ ] Clinical Edit

Data Sources:
- [ ] Only Administrative Databases
- [x] Databases + Prescriber-Supplied
Setting & Population

- Drug class for review: Anticonvulsants, Dravet Syndrome
- Age range: All appropriate MO HealthNet participants

Approval Criteria

- Documented compliance on current preferred therapy regimen OR
- Failure to achieve desired therapeutic outcomes with trial of 1 preferred agent
  - Documented trial period of preferred agents OR
  - Documented ADE/ADR to preferred agents AND
- For Epidiolex:
  - Documented diagnosis of Dravet syndrome, Lennox-Gastaut syndrome (LGS), or Tuberous sclerosis complex (TSC) AND
  - Trial of 2 or more anti-epileptic agents (includes clobazam, clonazepam, felbamate, lamotrigine, rufinamide, topiramate, valproate derivatives, or vigabatrin)
- For Diacomit:
  - Participant aged 2 years or older AND
  - Documented diagnosis of Dravet syndrome AND
  - Documented therapeutic trial (defined as 30 days) of both valproate and clobazam AND
  - Documentation of baseline seizure frequency and duration
- For Fintepla:
  - Participant aged 2 years or older AND
  - Documented diagnosis of Dravet syndrome or LGS AND
  - Documented therapeutic trial of Diacomit (defined as 30 days) AND
  - Documentation of baseline seizure frequency and duration AND
- Initial approval of prior authorization for Diacomit and Fintepla is 6 months, renewal of prior authorization may be up to 1 year with documentation of reduced seizure burden or improvement in quality of life using a validated scale for the disease state

Denial Criteria

- Lack of adequate trial on required preferred agents
- For Diacomit and Fintepla: documentation of moderate to severe hepatic or renal impairment
- For Fintepla: documented history of MAOI therapy in the past 45 days
- Therapy will be denied if all approval criteria are not met
- Claim exceeds quantity limitations:

<table>
<thead>
<tr>
<th>Drug Description</th>
<th>Generic Equivalent</th>
<th>Max Dosing Limitation</th>
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<tbody>
<tr>
<td>DIACOMIT 250 MG CAPSULE</td>
<td>STIRIPENTOL</td>
<td>12 capsules per day</td>
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<tr>
<td>DIACOMIT 250 MG POWDER</td>
<td>STIRIPENTOL</td>
<td>12 packets per day</td>
</tr>
<tr>
<td>DIACOMIT 500 MG CAPSULE</td>
<td>STIRIPENTOL</td>
<td>6 capsules per day</td>
</tr>
<tr>
<td>DIACOMIT 500 MG POWDER</td>
<td>STIRIPENTOL</td>
<td>6 packets per day</td>
</tr>
<tr>
<td>EPIDIOLEX 100 MG/ML</td>
<td>CANNABIDIOL</td>
<td>200 mL per fill</td>
</tr>
<tr>
<td>FINTEPLA 2.2 MG/ML</td>
<td>FENFLURAMINE</td>
<td>• With concomitant Diacomit: 17 mg per day</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Without concomitant Diacomit: 26 mg per day</td>
</tr>
</tbody>
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**Required Documentation**

<table>
<thead>
<tr>
<th>Laboratory Results:</th>
<th>Progress Notes:</th>
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<thead>
<tr>
<th>MedWatch Form:</th>
<th>Other:</th>
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**Disposition of Edit**

- Denial: Exception Code “0160” (Preferred Drug List)
- Rule Type: PDL

**Default Approval Period**

- 1 year

**References**