**Drug and Biologic Coverage Policy**

**Effective Date..... 7/1/2020**
**Next Review Date..... 7/1/2021**
**Coverage Policy Number ..... 2017**

# Selumetinib

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## Related Coverage Resources

***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. 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.*

## Coverage Policy

**Koselugo™ (selumetinib) is considered medically necessary when ALL of the following criteria are met:**

- ONE of the following:
  - Individual is 2 to 18 years of age
  - Individual is 19 years of age or older AND had initiated Koselugo therapy prior to age 19
- Treatment of Neurofibromatosis Type 1 (NF1)
- Documentation of symptomatic (for example, airway dysfunction, bladder/bowel dysfunction, disfigurement, motor dysfunction, pain, visual impairment), inoperable (for example, invasiveness, high vascularity, encasement of, or close proximity to, vital structures) plexiform neurofibromas

**Initial authorization is up to 12 months.**

**Koselugo (selumetinib) is considered medically necessary for continued use when there is documented beneficial clinical response (for example, stabilization in size of tumor or improvement in symptoms)**

**Reauthorization is up to 12 months.**

**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.**

**Koselugo (selumetinib) is considered experimental, investigational or unproven for ANY other use.**

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

## FDA Approved Indications

### FDA Approved Indication

Koselugo is a kinase inhibitor indicated for the treatment of pediatric patients 2 years of age and older with neurofibromatosis type 1 (NF1) who have symptomatic, inoperable plexiform neurofibromas (PN).

## Recommended Dosing

### FDA Recommended Dosing

***Refer to the prescribing information (product label) for complete dosing information. The following is from the "Highlights of Prescribing Information" section of the product label.*

The recommended dosage of Koselugo is 25 mg/m2 orally twice daily (approximately every 12 hours) until disease progression or unacceptable toxicity.
Take Koselugo on an empty stomach. Do not consume food 2 hours before each dose or 1 hour after each dose [see Clinical Pharmacology (12.3)]. The recommended dose of Koselugo based on body surface area (BSA) is shown in the table below.

**Recommended Dosage Based on Body Surface Area**

| Body Surface Area* | Recommended Dosage |
|---|---|
| 0.55 – 0.69 m2 | 20 mg in the morning and 10 mg in the evening |
| 0.70 – 0.89 m2 | 20 mg twice daily |
| 0.90 – 1.09 m2 | 25 mg twice daily |
| 1.10 – 1.29 m2 | 30 mg twice daily |
| 1.30 – 1.49 m2 | 35 mg twice daily |
| 1.50 – 1.69 m2 | 40 mg twice daily |
| 1.70 – 1.89 m2 | 45 mg twice daily |
| ≥ 1.90 m2 | 50 mg twice daily |

* The recommended dosage for patients with a BSA less than 0.55m2 has not been established.

## General Background

**Disease Overview**

Neurofibromatoses are a group of tumor suppressor syndromes that predisposes patients to an increased risk of nervous system tumors including neurofibromas, malignant peripheral nerve sheath tumors, and gliomas. Patients with neurofibromatosis type 1 (NF1) are also at increased risk for the development of gastrointestinal stromal tumor, breast cancer, leukemia, pheochromocytoma, duodenal carcinois tumor, and rhabdomyosarcoma. NF1 is the most common of the neurofibromatoses, occurring in approximately one in 2,500 to 3,000 individuals worldwide. NF1 is an autosomal dominate disorder, with 50% of children of affected parents

inheriting the mutated NF1 tumor-suppressor gene. However, up to 50% of the cases occur spontaneously in patients without a family history of NF1. (Cimino, 2018; Hirbe, 2014; Ly, 2019; Plotkin, 2018)

**The American Board of Internal Medicine's (ABIM) Foundation Choosing Wisely® Initiative:**
No recommendations are available for Neurofibromatosis.

**Other Covered Uses**
AHFS Drug Information 2020 Edition does not have a monograph for selumetinib (Koselugo).

**Compendium and Other Published Studies**
Compendia and other published clinical studies do not currently support any uses other than the FDA indication. Criteria will be updated as new published data are available.

## Coding/Billing Information

Note: Selumetinib is typically covered under pharmacy benefit plans. Certain prescription drugs require an authorization for coverage to ensure that appropriate treatment regimens are followed. Medical drug coding and diagnosis codes, however, are generally not required for pharmacy claims submissions, therefore, this section is not in use.

## References


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3. Koselugo™ capsules [prescribing information]. Wilmington, DE: AstraZeneca Pharmaceuticals; April 2020.
4. Ly KI, Blakeley JO. The diagnosis and management of neurofibromatosis type 1. Med Clin N Am. 2019; 103:1035-1054.
5. McEvoy GK, ed. AHFS 2020 Drug Information. Bethesda, MD: American Society of Health-Systems Pharmacists, Inc; 2020.
6. Plotkin SR, Wick A. Neurofibromatosis and Schwannomatosis. Semin Neurol. 2018; 38:73-85.
7. US National Institute of Health. In: ClinicalTrials.gov [Internet]. Bethesda (MD): National Library of Medicine (US). 2000- [cited 2020 March 23]. Available at: https://clinicaltrials.gov/ct2/results?cond=&term=selumetinib&cntry=&state=&city=&dist=. Search term: selumetinib.