MEDICAL MUTUAL®

Drug Policy

Policy:	Koselugo (selumetinib)	Annual Review Date: 05/18/2023 Last Revised Date: 05/18/2023

OVERVIEW

Koselugo is a MEK1/2 inhibitor indicated for the treatment of neurofibromatosis type 1 (NF1) in pediatric patients 2 years of age and older who have symptomatic, inoperable plexiform neurofibroma. The FDA granted Koselugo Priority Review, Breakthrough Therapy Designation, Rare Pediatric Disease Designation, and Orphan Drug Status.

POLICY STATEMENT

This policy involves the use of Koselugo. Prior authorization is recommended for pharmacy benefit coverage of Koselugo. Approval is recommended for those who meet the conditions of coverage in the **Criteria and Initial/Extended Approval** for the diagnosis provided. **Conditions Not Recommended for Approval** are listed following the recommended authorization criteria. Requests for uses not listed in this policy will be reviewed for evidence of efficacy and for medical necessity on a case-by-case basis.

Because of the specialized skills required for evaluation and diagnosis of patients treated with Koselugo as well as the monitoring required for adverse events and long-term efficacy, initial approval requires Koselugo be prescribed by or in consultation with a physician who specializes in the condition being treated. All approvals for initial therapy are provided for the initial approval duration noted below; if reauthorization is allowed, a response to therapy is required for continuation of therapy unless otherwise noted below.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Koselugo is recommended in those who meet the following criteria:

1. <u>Neurofibromatosis Type 1 (NF1), initial therapy</u>

Criteria. *Patient must meet the following criteria*

- **A.** The patient is between 2 and 18 years of age; AND
- **B.** The patient's BSA is greater than or equal to 0.55 m^2; AND
- **C.** The patient has at least one symptomatic, inoperable plexiform neurofibroma (PN), defined as a PN that cannot be completely removed without risk for substantial morbidity due to encasement of, or close proximity to, vital structures, invasiveness, or high vascularity or the PN; AND
- **D.** Koselugo is prescribed by, or in consultation with, a neurologist, oncologist, hematologist, or geneticist; AND
- **E.** The patient has a Karnofsky or Lansky performance level of greater than or equal to 70%; AND
- **F.** The patient has functional impairment or significant morbidity related to the target PN (e.g. motor dysfunction, airway dysfunction, bladder or bowel dysfunction, visual impairment, pain, disfigurement)

2. **Neurofibromatosis Type 1 (NF1), continuation of therapy**
Criteria. *Patient must meet the following criteria*

A. The patient is younger than 18 years of age; AND
B. Koselugo is prescribed by or in consultation with a neurologist, oncologist, hematologist, or geneticist; AND
C. The patient has not experienced disease progression while using Koselugo; AND
D. The patient has had a partial or complete response to therapy, as evidenced by at least one of the following: decrease in tumor volume, reduced tumor pain, improvement in physical functionality, increase in range of motion

3. **Recurrent or Progressive Circumscribed Glioma**
Criteria. *Patient must meet the following criteria*

A. Koselugo is prescribed by or in consultation with a hematologist or oncologist: AND
B. Koselugo is being used as single-agent therapy; AND
C. Tumors are positive for BRAF fusion or BRAF V600E activating mutation

4. **Langerhans Cell Histiocytosis**
Criteria. *Approve if Koselugo will be used as a single agent and is prescribed by or in consultation with a hematologist or oncologist.*

5. **Patients with another indication that is not listed but is cited in the National Comprehensive Cancer Network (NCCN) guidelines as a category 1, 2A, or 2B recommendation**
Criteria. *Prescriber will provide specific diagnosis for documentation. Approve.*

Initial Approval/ Extended Approval.
A) *Initial Approval:* 6 months
B) *Extended Approval:* 1 year

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Koselugo has not been shown to be effective, or there are limited or preliminary data or potential safety concerns that are not supportive of general approval for the following conditions. (Note: This is not an exhaustive list of Conditions Not Recommended for Approval).

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

Documentation Requirements:

The Company reserves the right to request additional documentation as part of its coverage determination process. The Company may deny reimbursement when it has determined that the drug provided or services performed were not medically necessary, investigational or experimental, not within the scope of benefits afforded to the member and/or a pattern of billing or other practice has been found to be either inappropriate or excessive. Additional documentation supporting medical necessity for the services provided must be made available upon request to the Company. Documentation requested may include patient records, test results and/or credentials of the provider ordering or performing a service. The Company also reserves the right to modify, revise, change, apply and interpret this policy at its sole discretion, and the exercise of this discretion shall be final and binding.

REFERENCES

1. Koselugo [prescribing information]. Wilmington, DE: AstraZeneca Pharmaceuticals LP; December 2021.
2. Selumetinib. In: DRUGDEX [online database]. Truven Health Analytics; Greenwood Village, CO. Last updated 13 March 2023. Accessed 16 May 2023.
3. Selumetinib. In: Lexi-Drugs. Lexicomp. Wolters Kluwer Clinical Drug Information, Inc.; Riverwoods, IL. Available at: http://www.online.lexi.com. Last updated 6 May 2023. Accessed 16 May 2023.
4. The NCCN Drugs & Biologics Compendium. © 2023 National Comprehensive Cancer Network. Available at: http://www.nccn.org. Accessed on 16 May 2023.