Koselugo (selumetinib)

Override(s)	Approval Duration
Prior Authorization Quantity Limit	1 year

Medications	Quantity Limit
Koselugo (selumetinib) capsules Koselugo (selumetinib) oral granules	May be subject to quantity limit

APPROVAL CRITERIA

Requests for Koselugo (selumetinib) or Koselugo (selumetinib) oral granules may be approved if the following criteria are met:

I. Individual is under 19 years of age;

OR

II. Individual has a diagnosis neurofibromatosis type 1 (NF1); **AND**
    A. Individual has symptomatic, inoperable plexiform neurofibromas (PN);

OR

III. Individual has a diagnosis of Langerhans Cell Histiocytosis (NCCN 2A); **AND**
    A. Individual is using in one of the following ways:
        1. Individual has mitogen-activated pathway (MAP) mutation, or no detectable mutation, or testing not available; **AND**
        2. Using for first-line or subsequent therapy; **AND**
        3. Using as a single agent;

        OR

        4. Individual is using for Langerhans Cell Histiocytosis (LCH)-associated abnormal CNS imaging/neurodegeneration (LACI/ND); **AND**
        5. Using for first-line or subsequent therapy; **AND**
        6. Using as a single agent;

OR

IV. Individual has a diagnosis of recurrent or progressive circumscribed astrocytic glioma (NCCN 2A); **AND**
    A. Individual is using as a single agent; **AND**
    B. Individual has one of the following disease types:
        1. Individual has a WHO grade 1 circumscribed glioma; **OR**
        2. Individual has a WHO grade 2 pleomorphic xanthoastrocytoma (PXA); **OR**
        3. Individual has a circumscribed glioma with a BRAF fusion or BRAF V600E mutation; **OR**
        4. Individual has a NF-1 mutated glioma;

AND

V. If request is for Koselugo (selumetinib) granules, individual meets roman numeral II., III., or IV., AND is unable to swallow the oral capsule dosage form due to a clinical condition including but not limited to the following:
   A. Dysphagia; **OR**
   B. Individual's age.

Requests for Koselugo (selumetinib) may not be approved for the following:

I. Individual has retinal vein occlusion (RVO) or retinal pigment epithelial detachment (RPED); **OR**
II. Individual has rhabdomyolysis.

Key References:

1. Awada G, Seremet T, Fostier K, Everaert H, Neyns B. Long-term disease control of Langerhans cell histiocytosis using combined BRAF and MEK inhibition. Blood Adv. 2018;2(16):2156-2158. doi:10.1182/bloodadvances.2018021782 Available at: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6113614/
2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: June 28, 2025
3. Dombi E, Baldwin A, Marcus LJ, et al. Activity of Selumetinib in Neurofibromatosis Type 1-Related Plexiform Neurofibromas. N Engl J Med. 2016;375(26):2550–2560. doi:10.1056/NEJMoa1605943.
4. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
5. Gross AM, Wolters PL, Dombi E, et al. Selumetinib in Children with Inoperable Plexiform Neurofibromas. N Engl J Med. 2020;382(15):1430–1442. doi:10.1056/NEJMoa1912735.
6. Janku F, Patel H, Raghavan VK, et al. MEK inhibition with trametinib in patients with non-Langerhans histiocytosis irrespective of BRAF mutation status. 2019 International ECD Medical Symposium 2019. Available at: https://erdheim-chester.org/wp-content/uploads/2019/10/Janku-Trametinib.pdf. Accessed June 28, 2025.
7. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2025; Updated periodically.
8. Lorillon G, Jouenne F, Baroudjian B, et al. Response to Trametinib of a Pulmonary Langerhans Cell Histiocytosis Harboring a MAP2K1 Deletion. Am J Respir Crit Care Med. 2018;198(5):675-678. doi:10.1164/rccm.201802-0275LE Available at: https://www.atsjournals.org/doi/pdf/10.1164/rccm.201802-0275LE. Accessed June 28, 2025.
9. NCCN Clinical Practice Guidelines in Oncology™. © 2025 National Comprehensive Cancer Network, Inc. For additional information visit the NCCN website: http://www.nccn.org/index.asp. Accessed on December 3, 2025.
   a. Central Nervous System Cancers. V2.2025. Revised August 28, 2025.
   b. Histiocytic Neoplasms. V2.2025. Revised November 21, 2025.