PHARMACY POLICY STATEMENT
Marketplace

DRUG NAME	Koselugo (selumetinib)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Koselugo, approved by the FDA in 2020, 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). It works by targeting mitogen-activated protein kinase kinases 1 and 2 (MEK1/2). MEK is a component of a pathway that is often activated in certain types of cancer.
NF1 is a rare, progressive genetic condition caused by a mutation in the *NF1* gene. histologically benign peripheral-nerve sheath tumors (PNST) that occur in up to 50% of NF1 patients. hat cannot be completely removed by surgery
transformation.
Koselugo was approved based on data from the phase 2 SPRINT clinical trial in which a majority of children had durable tumor shrinkage and clinical benefit such as pain reduction with treatment.

Koselugo (selumetinib) will be considered for coverage when the following criteria are met:

Neurofibromatosis Type 1 (NF1)

For **initial** authorization:

1. Member is at least 2 years of age; AND
2. Medication must be prescribed by or in consultation with a pediatric oncologist, neurologist, or geneticist; AND
3. Member has a confirmed diagnosis of neurofibromatosis type 1 (NF1) with at least 1 of the following:
   a) Positive genetic test for NF1
   b) 6 or more café-au-lait macules (CALMs)
   c) Axillary or inguinal freckling
   d) Optic glioma
   e) 2 or more Lisch nodules
   f) A distinctive osseous lesion
   g) First degree relative with NF1; AND
4. Member has at least one measurable plexiform neurofibromas (PN) as evidenced by MRI or PET-CT scan; AND
5. The plexiform neurofibromas (PN) is inoperable and cannot be removed completely by surgery without risk for substantial morbidity due to encasement of, or close proximity to, vital structures, invasiveness, or high vascularity of the PN; AND
6. Member has significant morbidity related to the PN (e.g., disfigurement, motor dysfunction, pain, airway dysfunction, visual impairment, bladder/bowel dysfunction).
7. **Dosage allowed/Quantity limit**: 25 mg/m^2 by mouth twice daily until disease progression or unacceptable toxicity. Capsules must be swallowed whole.
   (See Table 1 in prescribing information for recommended dosage based on body surface area).

   QL for 10 mg capsules: 224 per 28 days (8/day)
   QL for 25 mg capsules: 112 per 28 days (4/day)

If all the above requirements are met