Koselugo (selumetinib)
Policy Number: C19330-A

CRITERIA EFFECTIVE DATES:

ORIGINAL EFFECTIVE DATE	LAST REVIEWED DATE	NEXT REVIEW DUE BY OR BEFORE
07/23/2020	12/2/2020	12/2/2021
HCPCS CODING	**TYPE OF CRITERIA**	**LAST P&T APPROVAL/VERSION**
J8999 Prescription drug, oral, chemotherapeutic, NOC	RxPA	Q1 2021 20210127C19330-A

PRODUCTS AFFECTED:
Koselugo (selumetinib)

DRUG CLASS:
Antineoplastic - MEK Inhibitors

ROUTE OF ADMINISTRATION:
Oral

PLACE OF SERVICE:
Specialty Pharmacy
The recommendation is that medications in this policy will be for pharmacy benefit coverage and member self-administered

AVAILABLE DOSAGE FORMS:
Capsules: 10 mg and 25 mg

FDA-APPROVED USES:
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)

COMPENDIAL APPROVED OFF-LABELED USES:
None

COVERAGE CRITERIA: INITIAL AUTHORIZATION

DIAGNOSIS:
neurofibromatosis type 1 (NF1) plexiform neurofibromas (PN)

REQUIRED MEDICAL INFORMATION:
A. NEUROFIBROMATOSIS TYPE 1 PLEXIFORM NEUROFIBROMAS:
1. Documented diagnosis of NF1 plexiform neurofibromas (PN)
   AND
2. Documentation member has at least one measurable PN, defined as a lesion ≥ 3 cm measured in one dimension
3. Prescriber attests that complete resection of PN is not considered to be feasible without

substantial risk or morbidity (e.g., due to encasement of, or close proximity to, vital structures, invasiveness, or high vascularity of the PN
AND

4. Prescriber attests that member is able to swallow capsules whole and take medication on an empty stomach including 2 hours prior to administration and 1 hour after
AND
5. Prescriber attests to counseling member to refrain from taking vitamin-E preparations and other medications that can cause drug-drug interactions
AND
6. Documentation that member is symptomatic (i.e. pain, motor dysfunction, and visual loss)
AND
7. Prescriber attests that baseline ophthalmic assessment has been done and prescriber agrees to monitor for ocular toxicities AND baseline left ventricular ejection fraction (LVEF) has been assessed and prescriber agrees to monitor LVEF regularly throughout treatment with Koselugo.
AND
8. Prescriber attests to counseling of females of reproductive potential only: member will be advised to use effective contraception during treatment with Koselugo and for 1 week after the last dose OR Males with female partners of reproductive potential only: member will be advised to use effective contraception during treatment with Koselugo and for 1 week after the last dose

DURATION OF APPROVAL:
Initial authorization: 6 months**,** Continuation of Therapy: 12 months

QUANTITY:
25mg/m2 twice a day
120 capsules per 30 days of either strength (10mg or 25mg)

PRESCRIBER REQUIREMENTS:
Prescribed by or in consultation with an oncologist or a neurologist (consultation notes should be provided as documentation at a minimum of annually)

AGE RESTRICTIONS:
≥2 years with a BSA of ≥0.55 m2 (able to swallow whole capsule)

CONTINUATION OF THERAPY:
A. NEUROFIBROMATOSIS TYPE 1 PLEXIFORM NEUROFIBROMAS:

1. Adherence to therapy at least 85% of the time as verified by Prescriber and member's medication fill history (review Rx history for compliance)- unless therapy held for toxicity
AND
2. Documentation of no evidence of disease progression or unacceptable toxicity
AND
3. Prescriber attests to continued counseling of females of reproductive potential only: member will be advised to use effective contraception during treatment with Koselugo and for 1 week after the last dose OR Males with female partners of reproductive potential only: member will be advised to use effective contraception during treatment with Koselugo and for 1 week after the last dose
AND
4. Prescriber attests that member continues to be able to swallow capsules whole and take

medication on an empty stomach including 2 hours prior to administration and 1 hour after

AND

5. Prescriber attest that to monitoring for ocular toxicities AND baseline left ventricular ejection fraction (LVEF) has been assessed and will continue

CONTRAINDICATIONS/EXCLUSIONS/DISCONTINUATION:
All other uses of Koselugo (selumetinib) are considered experimental/investigational and therefore, will follow Molina's Off- Label policy.

OTHER SPECIAL CONSIDERATIONS:
None

BACKGROUND:
Neurofibromatosis type 1 (NF1) is a rare, progressive condition caused by a mutation or flaw in the NF1 gene. It occurs in approximately 1 out of 2,600–3,000 infants. About 50% of cases are due to an inherited autosomal dominant genetic disorder, while the other 50% of cases are due to sporadic genetic mutations. Plexiform neurofibromas (PN) are tumors involving the nerve sheaths which can grow anywhere in the body, including the face, extremities, areas around the spine and deep in the body where they may affect organs. NF1 is usually diagnosed in early childhood. It is characterized by changes in skin pigmentation, neurologic and skeletal impairments, and risk for development of benign and malignant tumors throughout life. The risk of developing a cancer is estimated to be about 7%. Between 30% and 50% of patients born with NF1 develop one or more PNs. An Italian study by Maria Masocco in the Orphanet Journal of Rare Diseases (2011) noted the mean age for NF1-associated death was approximately 20 years lower than that for the general population.

APPENDIX:
None

Documentation Requirements:
Molina Healthcare reserves the right to require that additional documentation be made available as part of its coverage determination; quality improvement; and fraud; waste and abuse prevention processes. Documentation required may include, but is not limited to, member records, test results and credentials of the provider ordering or performing a drug or service. Molina Healthcare may deny reimbursement or take additional appropriate action if the documentation provided does not support the initial determination that the drugs or services were medically necessary, not investigational or experimental, and otherwise within the scope of benefits afforded to the member, and/or the documentation demonstrates a pattern of billing or other practice that is inappropriate or excessive.

REFERENCES:

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3. Gross AM, Wolters P, Baldwin A, et al. SPRINT: Phase II study of the MEK 1/2 inhibitor

selumetinib (AZD6244, ARRY-142886) in children with neurofibromatosis type 1 (NF1) and inoperable plexiform neurofibromas (PN) [Abstract]. J Clin Oncol 2018; 36(Suppl15):10503.
4. Dombi, et al. Activity of Selumetinib in Neurofibromatosis Type 1–Related Plexiform Neurofibromas. NEJM. 2016;375:2550–2560. doi: 10.1056/NEJMoa1605943
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6. National Institutes of Health. In NIH trial, selumetinib shrinks tumors, provides clinical benefit for children with NF1. News Release; www.nih.gov. Published March 18, 2020. Accessed April 17, 2020.
7. Masocco, et al. Mortality associated with neurofibromatosis type 1: A study based on Italian death certificates (1995-2006). Orphanet J Rare Dis. 2011;6:11. doi: 10.1186/1750-1172-6-11
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