Koselugo®
(selumetinib)
10 mg & 25 mg capsules

KOSELUGO CODING RESOURCE

The Koselugo Coding Resource is intended to provide objective and publicly available coding and billing information for non-contracted specialty pharmacies, retail pharmacies, hospitals, and outpatient clinics seeking to dispense Koselugo.

This document is provided for informational purposes only and is not legal advice or official guidance from payers. It is not intended to increase or maximize reimbursement by any payer. Alexion does not warrant, promise, guarantee, or make any statement that the use of this information will result in coverage or payment for Koselugo or that any payment received will cover providers' costs. Alexion is not responsible for any action providers take in billing for, or appealing, Koselugo claims.

Hospitals and physicians are responsible for compliance with Medicare and other payer rules and requirements and for the information submitted with all claims and appeals. Before any claims or appeals are submitted, hospitals and physicians should review official payer instructions and requirements, should confirm the accuracy of their coding or billing practices with these payers, and should use independent judgment when selecting codes that most appropriately describe the services or supplies provided to a patient.

Please visit **www.koselugohcp.com** for additional information. For inquiries regarding reimbursement, please call 1-888-765-4747 to speak with a OneSource™ patient support specialist who can connect you with your local Field Reimbursement Manager. OneSource™ is available Monday through Friday, 8:30 AM–8:00 PM ET.

INDICATION

Koselugo is 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).

Please see Important Safety Information on pages 3 and 4 and the accompanying full Prescribing Information for KOSELUGO® (selumetinib).

Product Overview[1]

KOSELUGO® (selumetinib) is the first and only FDA-approved prescription medicine that is used to treat pediatric patients 2 years of age and older with neurofibromatosis type 1 (NF1) who have plexiform neurofibromas that cannot be completely removed by surgery.

Koselugo is available as oral capsules taken twice daily on an empty stomach. Dosing is individualized based on each patient's body surface area (up to a maximum single dose of 50 mg).

Koselugo is available in 10 mg and 25 mg capsules and supplied in 28- and 60-count bottles.

National Drug Code (NDC)

Some payers may require drugs like Koselugo to be billed on pharmacy claims with the product's NDC. The following NDCs are used to identify Koselugo when submitting a pharmacy claim:

10-digit NDC[1]

Dosage	Bottle Count	Code
10 mg Capsules	28	0310-0610-28
25 mg Capsules	28	0310-0625-28
10 mg Capsules	60	0310-0610-60
25 mg Capsules	60	0310-0625-60

11-digit NDC[1]

Dosage	Bottle Count	Code
10 mg Capsules	28	00310-0610-28
25 mg Capsules	28	00310-0625-28
10 mg Capsules	60	00310-0610-60
25 mg Capsules	60	00310-0625-60

Payers typically require healthcare professionals to use the Health Insurance Portability and Accountability Act (HIPAA)-compliant, 11-digit NDC format. Please note that payers have different guidance for placement of the NDC on pharmacy claims. Typically, the 11-digit NDC is reported without any dashes or other punctuation.[2]

Diagnosis Codes[3]

To describe a patient diagnosed with neurofibromatosis, type 1, the following International Classification of Diseases, 10th Revision, Clinical Modification (ICD-10-CM) codes may be appropriate to indicate the patient's condition:

ICD-10-CM	Description
Q85.01	Neurofibromatosis, type 1
D33.3	Benign neoplasm of cranial nerves
D36.10	Benign neoplasm of peripheral nerves and autonomic nervous system, unspecified
D36.11	Benign neoplasm of peripheral nerves and autonomic nervous system of face, head, and neck
D36.12	Benign neoplasm of peripheral nerves and autonomic nervous system, upper limb, including shoulder
D36.13	Benign neoplasm of peripheral nerves and autonomic nervous system of lower limb, including hip
D36.14	Benign neoplasm of peripheral nerves and autonomic nervous system of thorax
D36.15	Benign neoplasm of peripheral nerves and autonomic nervous system of abdomen
D36.16	Benign neoplasm of peripheral nerves and autonomic nervous system of pelvis
D36.17	Benign neoplasm of peripheral nerves and autonomic nervous system of trunk, unspecified

Please see Important Safety Information on pages 3 and 4 and the accompanying full Prescribing Information for KOSELUGO® (selumetinib).

INDICATION AND SELECT IMPORTANT SAFETY INFORMATION for KOSELUGO® (selumetinib)

INDICATION

Koselugo is 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).

SELECT IMPORTANT SAFETY INFORMATION

Cardiomyopathy. A decrease in left ventricular ejection fraction (LVEF) ≥10% below baseline occurred in 23% of 74 pediatric patients who received Koselugo in SPRINT. Four percent of patients experienced decreased LVEF below the institutional lower limit of normal (LLN). Grade 3 decreased LVEF occurred in one patient and resulted in dose reduction. All patients with decreased LVEF were asymptomatic and identified during routine echocardiography. Decreased LVEF resolved in 71% of these patients. Decreased LVEF resulting in permanent discontinuation of Koselugo occurred in a pediatric population with NF1 in an expanded access program. The safety of Koselugo has not been established in patients with a history of impaired LVEF or a baseline ejection fraction that is below the institutional LLN.

Assess ejection fraction by echocardiogram prior to initiating treatment, every 3 months during the first year of treatment, every 6 months thereafter, and as clinically indicated. Withhold, reduce dose, or permanently discontinue Koselugo based on severity of adverse reaction. In patients who interrupt Koselugo for decreased LVEF, obtain an echocardiogram or a cardiac MRI every 3 to 6 weeks. Upon resolution of decreased LVEF, obtain an echocardiogram or a cardiac MRI every 2 to 3 months.

Ocular Toxicity. Blurred vision, photophobia, cataracts, and ocular hypertension occurred in 15% of 74 pediatric patients receiving Koselugo in SPRINT. Blurred vision resulted in dose interruption in 2.7% of patients. Ocular toxicity resolved in 82% of 11 patients. Retinal pigment epithelial detachment (RPED) occurred in the pediatric population during treatment with single agent Koselugo and resulted in permanent discontinuation.

Conduct ophthalmic assessments prior to initiating Koselugo, at regular intervals during treatment, and for new or worsening visual changes. Permanently discontinue Koselugo in patients with retinal vein occlusion (RVO). Withhold Koselugo in patients with RPED, conduct ophthalmic assessments every 3 weeks until resolution, and resume Koselugo at a reduced dose. For other ocular toxicities, withhold, reduce dose, or permanently discontinue Koselugo based on severity of adverse reaction.

Gastrointestinal Toxicity. Diarrhea occurred in 77% of 74 pediatric patients who received Koselugo in SPRINT, including Grade 3 in 15% of patients. Diarrhea resulting in permanent discontinuation occurred in 1.4% of patients. Diarrhea resulting in dose interruption or dose reduction occurred in 15% and 1.4% of patients, respectively. The median time to first onset of diarrhea was 17 days, and the median duration was 2 days.

Advise patients to start an anti-diarrheal agent (eg, loperamide) and to increase fluid intake immediately after the first episode of diarrhea. Withhold, reduce dose, or permanently discontinue Koselugo based on severity of adverse reaction.

Skin Toxicity. Rash occurred in 91% of 74 pediatric patients who received Koselugo in SPRINT. The most frequent rashes included dermatitis acneiform (54%), maculopapular rash (39%), and eczema (28%). Grade 3 rash occurred in 8% of patients. Rash resulted in dose interruption in 11% of patients and dose reduction in 4% of patients. Monitor for severe skin rashes. Withhold, reduce dose, or permanently discontinue Koselugo based on severity of adverse reaction.

Increased Creatine Phosphokinase (CPK). Increased CPK occurred in 76% of 74 pediatric patients who received Koselugo in SPRINT, including Grade 3 or 4 in 9% of patients. Increased CPK resulted in dose reduction in 7% of patients. Increased CPK concurrent with myalgia occurred in 8% of patients, including one patient who permanently discontinued Koselugo for myalgia.

Obtain serum CPK prior to initiating Koselugo, periodically during treatment, and as clinically indicated. If increased CPK occurs, evaluate patients for rhabdomyolysis or other causes. Withhold, reduce dose, or permanently discontinue Koselugo based on severity of adverse reaction.

Please see Important Safety Information on pages 3 and 4 and the accompanying full Prescribing Information for KOSELUGO® (selumetinib).

SELECT IMPORTANT SAFETY INFORMATION for KOSELUGO® (selumetinib) (cont.)

Increased Levels of Vitamin E and Risk of Bleeding. Koselugo capsules contain vitamin E (10 mg capsules contain 32 mg vitamin E as the excipient, D-alpha-tocopheryl polyethylene glycol 1000 succinate [TPGS], while Koselugo 25 mg capsules contain 36 mg vitamin E as TPGS). Vitamin E can inhibit platelet aggregation and antagonize vitamin K-dependent clotting factors. Daily vitamin E intake that exceeds the recommended or safe limits may increase the risk of bleeding. Supplemental vitamin E is not recommended if daily vitamin E intake (including the amount of vitamin E in Koselugo and supplement) will exceed the recommended or safe limits.

An increased risk of bleeding may occur in patients who are coadministered vitamin-K antagonists or anti-platelet antagonists with Koselugo. Monitor for bleeding in these patients and increase international normalized ratio (INR) monitoring in patients taking a vitamin-K antagonist. Perform anticoagulant assessments more frequently and adjust the dose of vitamin K antagonists or anti-platelet agents as appropriate.

Embryo-Fetal Toxicity. Based on findings from animal studies, Koselugo can cause fetal harm when administered to a pregnant woman. In animal studies, administration of selumetinib to mice during organogenesis caused reduced fetal weight, adverse structural defects, and effects on embryo-fetal survival at approximate exposures >5 times the human exposure at the clinical dose of 25 mg/m^2 twice daily. Advise pregnant women of the potential risk to a fetus. Advise females of reproductive potential to use effective contraception during treatment with Koselugo and for 1 week after the last dose. Advise males with female partners of reproductive potential to use effective contraception during treatment with Koselugo and for 1 week after the last dose.

Breastfeeding. Due to the potential for adverse reactions in a breastfed child, advise women not to breastfeed during treatment with Koselugo and for 1 week after the last dose.

Concomitant use of Koselugo with a strong or moderate CYP3A4 inhibitor or fluconazole increased selumetinib plasma concentrations, which may increase the risk of adverse reactions. Avoid coadministration of strong or moderate CYP3A4 inhibitors or fluconazole with Koselugo. If coadministration with strong or moderate CYP3A4 inhibitors or fluconazole cannot be avoided, reduce Koselugo dosage.

Concomitant use of Koselugo with a strong or moderate CYP3A4 inducer decreased selumetinib plasma concentrations, which may reduce Koselugo efficacy. Avoid concomitant use of strong or moderate CYP3A4 inducers with Koselugo.

The most common adverse reactions ≥40% are: vomiting, rash (all), abdominal pain, diarrhea, nausea, dry skin, musculoskeletal pain, fatigue, pyrexia, acneiform rash, stomatitis, headache, paronychia, and pruritus.

Please see Important Safety Information on pages 3 and 4 and the accompanying full Prescribing Information for KOSELUGO® (selumetinib).

OneSource™ Offers Patient Support

Contact the Alexion OneSource™ Team at:

Phone:
1-888-765-4747
Monday to Friday, 8:30 AM to 8:00 PM ET

Online:
AlexionOneSource.com

References: 1. KOSELUGO. Prescribing information. AstraZeneca Pharmaceuticals LP. **2.** Future format of the National Drug Code; public hearing; request for comments. *Fed Regist*. 2018;83;152:38666-38668. Accessed April 20, 2022. https://www.federalregister.gov/documents/2018/08/07/2018-16807/future-format-of-thenational-drug-code-public-hearing-request-for-comments **3.** Centers for Medicare & Medicaid Services. 2022 ICD-10-CM. Updated February 1, 2022. Accessed April 20, 2022. https://www.cms.gov/medicare/icd-10/2022-icd-10-cm

US/KOS-NF1/0385 05/22

ALEXION®
AstraZeneca Rare Disease