BlueCross BlueShield Arizona

An Independent Licensee of the Blue Cross Blue Shield Association

PHARMACY COVERAGE GUIDELINE

OJJAARA® (momelotinib)
Generic Equivalent (if available)

This Pharmacy Coverage Guideline (PCG):
- Provides information about the reasons, basis, and information sources we use for coverage decisions
- Is not an opinion that a drug (collectively "Service") is clinically appropriate or inappropriate for a patient
- Is not a substitute for a provider's judgment (Provider and patient are responsible for all decisions about appropriateness of care)
- Is subject to all provisions e.g. (benefit coverage, limits, and exclusions) in the member's benefit plan; and
- Is subject to change as new information becomes available.

Scope
- This PCG applies to Commercial and Marketplace plans
- This PCG does not apply to the Federal Employee Program, Medicare Advantage, Medicaid or members of out-of-state Blue Cross and/or Blue Shield Plans

Instructions & Guidance
- To determine whether a member is eligible for the Service, read the entire PCG.
- This PCG is used for FDA approved indications including, but not limited to, a diagnosis and/or treatment with dosing, frequency, and duration.
- Use of a drug outside the FDA approved guidelines, refer to the appropriate Off-Label Use policy.
- The "Criteria" section outlines the factors and information we use to decide if the Service is medically necessary as defined in the Member's benefit plan.
- The "Description" section describes the Service.
- The "Definition" section defines certain words, terms or items within the policy and may include tables and charts.
- The "Resources" section lists the information and materials we considered in developing this PCG
- **We do not accept patient use of samples as evidence of an initial course of treatment, justification for continuation of therapy, or evidence of adequate trial and failure.**
- Information about medications that require prior authorization is available at www.azblue.com/pharmacy. You must fully complete the request form and provide chart notes, lab workup and any other supporting documentation. The prescribing provider must sign the form. Fax the form to BCBSAZ Pharmacy Management at (602) 864-3126 or email it to Pharmacyprecert@azblue.com.

Criteria:

➢ **Criteria for initial therapy:** Ojjaara (momelotinib) and/or generic equivalent (if available) is considered ***medically necessary*** and will be approved when **ALL** the following criteria are met:

1. Prescriber is a physician specializing in the patient's diagnosis or is in consultation with an Oncologist

2. Individual is 18 years of age or older

3. Individual has a confirmed diagnosis of **ONE** of the following:
    a. Intermediate or high-risk myelofibrosis (MF), including primary MF or secondary MF [post-polycythemia vera (PV) and post-essential thrombocythemia (ET)] as defined by the Dynamic International Prognostic Scoring System (DIPSS) or International Prognostic Scoring System (IPSS) for MF (see Definitions section)

BLUE CROSS®, BLUE SHIELD® and the Cross and Shield Symbols are registered service marks of the Blue Cross and Blue Shield Association, an association of independent Blue Cross and Blue Shield Plans. All other trademarks and service marks contained in this guideline are the property of their respective owners, which are not affiliated with BCBSAZ.

b. Other request for a specific oncologic direct treatment use that is found and listed in the National Comprehensive Cancer Network (NCCN) Guidelines with Categories of Evidence and Consensus of 1 and 2A

4. Individual has symptomatic anemia (hemoglobin is less than 10 g/dL)

5. Individual has received and completed **ALL** the following **baseline tests** before initiation of treatment and with continued monitoring of the individual as clinically appropriate:
   a. Complete blood count (CBC) with platelets
   b. Liver function tests
   c. For an individual with HBV infection, hepatitis B serologies

6. **If available:** Individual has failure after adequate trial, contraindication per FDA label, intolerance, or is not a candidate for a **generic equivalent** [Note: Failure, contraindication or intolerance to the generic should be reported to the FDA] (see Definitions section)

7. Individual has documented failure, contraindication per FDA label, intolerance, or is not a candidate for Jakafi (ruxolitinib)

8. Will not be used with other Tyrosine Kinase Inhibitors or other Janus Associated Kinase Inhibitors (such as Xeljanz (tofacitinib), Xeljanz (tofacitinib) XR, Olumiant (baricitinib), Rinvoq (upadactinib), Inrebic (fedratinib), Vonjo (pacritinib) or others)

9. Individual does not have an active infection (e.g., bacterial, viral)

10. Individual does not have end stage renal disease receiving dialysis

Initial approval duration: 6 months

- **Criteria for continuation of coverage (renewal request):** Ojjaara (momelotinib) and/or generic equivalent (if available) is considered ***medically necessary*** and will be approved when **ALL** the following criteria are met **(samples are not considered for continuation of therapy)**:

1. Individual continues to be seen by a physician specializing in the patient's diagnosis or is in consultation with an Oncologist

2. Individual's condition has responded while on therapy with response defined as there is documented evidence of efficacy, disease stability and/or improvement

3. Individual has been adherent with the medication

4. Requested dose is at least 100 mg once daily

5. **If available:** Individual has failure after adequate trial, contraindication per FDA label, intolerance, or is not a candidate for a **generic equivalent** [Note: Failure, contraindication or intolerance to the generic should be reported to the FDA] (see Definitions section)

BLUE CROSS®, BLUE SHIELD® and the Cross and Shield Symbols are registered service marks of the Blue Cross and Blue Shield Association, an association of independent Blue Cross and Blue Shield Plans. All other trademarks and service marks contained in this guideline are the property of their respective owners, which are not affiliated with BCBSAZ.

6. Individual has not developed any contraindications or other significant adverse drug effects that may exclude continued use such as:
    a. A recurrence of ALT or AST elevations >5 × ULN
    b. Serious cardiovascular events such as myocardial infarction, stroke
    c. Deep venous thrombosis, pulmonary embolism, and arterial thrombosis
    d. Lymphoma and other malignancies excluding nonmelanoma skin cancer (NMSC)

7. Will not be used with other Tyrosine Kinase Inhibitors or other Janus Associated Kinase Inhibitors (such as Xeljanz (tofacitinib), Xeljanz (tofacitinib) XR, Olumiant (baricitinib), Rinvoq (upadactinib), Inrebic (fedratinib), Vonjo (pacritinib) or others)

8. Individual does not have an active infection (e.g., bacterial, viral)

9. Individual does not have end stage renal disease receiving dialysis

Renewal duration: 12 months

- Criteria for a request for non-FDA use or indication, treatment with dosing, frequency, or duration outside the FDA-approved dosing, frequency, and duration, refer to one of the following Pharmacy Coverage Guideline:

    1. **Off-Label Use of Non-Cancer Medications**

    2. **Off-Label Use of Cancer Medications**

Description:

Ojjaara (momelotinib) is a kinase inhibitor indicated for the treatment of intermediate or high-risk myelofibrosis (MF), including primary MF or secondary MF [post-polycythemia vera (PV) and post-essential thrombocythemia (ET)], in adults with anemia. Momelotinib is an inhibitor of wild type Janus Kinase 1 and 2 (JAK1/JAK2) and mutant JAK2V617F, which contribute to signaling of a number of cytokines and growth factors that are important for hematopoiesis and immune function.

MF, a Philadelphia chromosome-negative chronic myeloproliferative disorder, is characterized by progressive anemia, bone marrow fibrosis, splenomegaly and constitutional symptoms. Up to 30% of patients are initially asymptomatic. Many patients present with symptoms from anemia, splenomegaly or constitutional symptoms (severe fatigue, low grade fever, pruritus, night sweats and weight loss). As the disease evolves, all patients become symptomatic due to marrow failure and increasing splenomegaly resulting in abdominal symptoms and early satiety.

The International Working Group (IWG) consensus for Myelofibrosis Research and Treatment has devised an international prognostic scoring system (IPSS) that uses presenting signs and symptoms to assign risk categories. Individuals with zero (low risk), one (intermediate risk-1), two (intermediate risk-2), or ≥3 (high risk) at presentation had non-overlapping median survivals of 135, 95, 48, and 27 months, respectively. The following five adverse prognostic features were noted by the IWP IPSS: age > 65 years; presence of constitutional symptoms

BLUE CROSS®, BLUE SHIELD® and the Cross and Shield Symbols are registered service marks of the Blue Cross and Blue Shield Association, an association of independent Blue Cross and Blue Shield Plans. All other trademarks and service marks contained in this guideline are the property of their respective owners, which are not affiliated with BCBSAZ.

(weight loss >10 % from baseline, night sweats, or unexplained fever); hemoglobin <10 g/dL; leukocyte count > 25 X 10^9/L; and circulating blast cells ≥ 1%.

PV is a chronic myeloproliferative disorder that causes the bone marrow to produce too many red blood cells. The median age at presentation is 60 years. Patients often present with either arterial or venous vascular occlusive events. The events are predominantly coronary and cerebral but can involve the skin and gastrointestinal tract. Over time PV may evolve to MF, acute myeloid leukemia (AML), or myelodysplastic syndrome (MDS). The mainstay of therapy for PV is phlebotomy which removes excess red blood cells and lowers blood viscosity. In general, the goal of phlebotomy is to keep the hematocrit below 45% in men and 42% in women.

Definitions:

U.S. Food and Drug Administration (FDA) MedWatch Forms for FDA Safety Reporting
MedWatch Forms for FDA Safety Reporting | FDA

ECOG Performance status:

Eastern Co-operative Oncology Group (ECOG) Performance Status	
Grade	ECOG description
0	Fully active, able to carry on all pre-disease performance without restriction
1	Restricted in physically strenuous activity but ambulatory and able to carry out work of a light or sedentary nature, e.g., light housework, office work
2	Ambulatory and capable of all selfcare but unable to carry out any work activities. Up and about more than 50% of waking hours
3	Capable of only limited selfcare, confined to bed or chair more than 50% of waking hours
4	Completely disabled. Cannot carry on any self-care. Totally confined to bed or chair
5	Dead
Oken, M.M., Creech, R.H., Tormey, D.C., Horton, J., Davis, T.E., McFadden, E.T., Carbone, P.P.: Toxicity And Response Criteria Of The Eastern Cooperative Oncology Group. Am J Clin Oncol 5:649-655, 1982	

NCCN recommendation definitions:

Category 1:
Based upon high-level evidence, there is uniform NCCN consensus that the intervention is appropriate.

Category 2A:
Based upon lower-level evidence, there is uniform NCCN consensus that the intervention is appropriate.

Category 2B:
Based upon lower-level evidence, there is NCCN consensus that the intervention is appropriate.

Category 3:
Based upon any level of evidence, there is major NCCN disagreement that the intervention is appropriate

Myelofibrosis:

These risk stratification systems have been studied and validated only in patient with PMF, but clinically have been used for stratification of patients with Post-PV MF or Post-ET MF. Novel prognostic models are being developed for risk stratification of patients with Post-PV MF or Post-ET MF. IPSS should be used at time of diagnosis, DIPSS-PLUS is preferred during the course of treatment, DIPSS can be used if karyotyping is not available

BLUE CROSS®, BLUE SHIELD® and the Cross and Shield Symbols are registered service marks of the Blue Cross and Blue Shield Association, an association of independent Blue Cross and Blue Shield Plans. All other trademarks and service marks contained in this guideline are the property of their respective owners, which are not affiliated with BCBSAZ.

PHARMACY COVERAGE GUIDELINE

OJJAARA® (momelotinib)
Generic Equivalent (if available)

International Working Group (IWG) International prognostic scoring system (IPSS):

Risk Stratification for Myelofibrosis (IPSS)	
	Points
Age > 65 years	1
Constitutional symptoms: Weight loss > 10 % from baseline Night sweats Unexplained fever	1
Hemoglobin <10 g/dL	1
Leukocyte count > 25 X 10^9/L	1
Circulating blast cells ≥ 1%	1
Risk Group	
Low risk	0 points
Intermediate risk-1	1 point
Intermediate risk-2	2 points
High risk	3 or more points

Dynamic International Prognostic System (DIPSS):

Prognostic Variable	Points		
	0	1	2
Age (y)	≤ 65	> 65	
Constitutional symptoms (Y/N)	N	Y	
Hemoglobin (g/dL)	≥ 10		< 10
WBC (x 10^9/L)	≤ 25	> 25	
Peripheral blood blasts (%)	< 1	≥ 1	
Risk Group	Points		
Low	0		
Intermediate-1	1 or 2		
Intermediate-2	3 or 4		
High	5 or 6		

Dynamic International Prognostic System Plus (DIPSS-Plus):

Prognostic Variable	Points
DIPSS low risk	0
DIPSS Intermediate-1	1
DIPSS Intermediate-2	2
DIPSS high risk	3

BLUE CROSS®, BLUE SHIELD® and the Cross and Shield Symbols are registered service marks of the Blue Cross and Blue Shield Association, an association of independent Blue Cross and Blue Shield Plans. All other trademarks and service marks contained in this guideline are the property of their respective owners, which are not affiliated with BCBSAZ.

Platelets < 100 x 10^9/L	1
Transfusion need	1
Unfavorable karyotype*	1
Risk Group	Points
Low	0
Intermediate-1	1
Intermediate-2	2 or 3
High	4 to 6
*Unfavorable karyotype: complex karyotype or sole or two abnormalities that include trisomy 8, 7/7q, i(17q), 5/5q-, 12p-, inv(3), or 11q23 rearrangement	

Assessment of Symptom Burden:

MPN-SAF is recommended for assessment at baseline and MPN-SAF TSS is recommended for monitoring during the course of treatment

Myeloproliferative Neoplasm Symptom Assessment Form (MPN-SAF)		
	Circle the one number that describes, **during the past week**, how much difficulty you had with each of the following symptoms	
Early satiety	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Abdominal pain	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Abdominal discomfort	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Inactivity	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Problems with headaches	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Problems with concentration compared to before Dx	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Dizziness/vertigo/lightheaded	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Numbness tingling hands/feet	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Difficulty sleeping	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Depressed or sad mood	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Problems with sexual desire or ability	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Cough	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Night sweats	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Itching	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Bone pain – not joint pain or arthritis	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Unintentional weight loss	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Fever	Absent = 0; Daily = 10	0-1-2-3-4-5-6-7-8-9-10
Overall quality of life	As good as it can be = 0; As bad as it can be = 10	0-1-2-3-4-5-6-7-8-9-10

BLUE CROSS®, BLUE SHIELD® and the Cross and Shield Symbols are registered service marks of the Blue Cross and Blue Shield Association, an association of independent Blue Cross and Blue Shield Plans. All other trademarks and service marks contained in this guideline are the property of their respective owners, which are not affiliated with BCBSAZ.

PHARMACY COVERAGE GUIDELINE

OJJAARA® (momelotinib)
Generic Equivalent (if available)

Myeloproliferative Neoplasm Symptom Assessment Form Total Symptom Score (MPN-SAF TSS; MPN 10)		
Rate fatigue (weariness, tiredness) that describes your worst level of fatigue **during the past 24 hours**	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Circle the one number that describes, **during the past week**, how much difficulty you had with each of the following symptoms		
Early satiety	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Abdominal discomfort	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Inactivity	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Problems with concentration compared to before Dx	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Numbness tingling hands/feet	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Night sweats	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Itching	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Bone pain – not joint pain or arthritis	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Unintentional weight loss	Absent = 0; Worst imaginable = 10	0-1-2-3-4-5-6-7-8-9-10
Fever	Absent = 0; Daily = 10	0-1-2-3-4-5-6-7-8-9-10

Polycythemia vera:

Low-risk patients
- Age < 60 years
- No history of thrombosis

High-risk patients:
- Age ≥ 60 years
- History of thrombosis

Potential indications for cytoreductive therapy:
- New thrombosis or disease related major bleeding
- Frequent and/or persistent need for phlebotomy, but with poor tolerance for phlebotomy
- Splenomegaly
- Thrombocytosis
- Leukocytosis
- Disease related symptoms (e.g., pruritus, night sweats, fatigue)

Resources:

Ojjaara (momelotinib) product information, revised by GlaxoSmithKline LLC. 09-2023. Available at DailyMed http://dailymed.nlm.nih.gov. Accessed October 15, 2024.

National Comprehensive Cancer Network (NCCN) Clinical Practice Guidelines in Oncology (NCCN Guidelines®): Myeloproliferative Neoplasms Version 2.2024 –Updated August 08, 2024. Available at https://www.nccn.org. Accessed October 15, 2024.

Off Label Use of Cancer Medications: A.R.S. §§ 20-826(R) & (S). Subscription contracts; definitions.

Off Label Use of Cancer Medications: A.R.S. §§ 20-1057(V) & (W). Evidence of coverage by health care service organizations; renewability; definitions.

ORIGINAL EFFECTIVE DATE: 11/16/2023 | ARCHIVE DATE: | LAST REVIEW DATE: 11/21/2024 | LAST CRITERIA REVISION DATE: 11/21/2024

BLUE CROSS®, BLUE SHIELD® and the Cross and Shield Symbols are registered service marks of the Blue Cross and Blue Shield Association, an association of independent Blue Cross and Blue Shield Plans. All other trademarks and service marks contained in this guideline are the property of their respective owners, which are not affiliated with BCBSAZ.