AETNA BETTER HEALTH®
Coverage Policy/Guideline

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Effective Date:	8/10/2023	Last Review Date:	5/31/2023

Applies to:	☐Illinois	☐Florida	☐Florida Kids
	☒New Jersey	☒Maryland	☒Michigan
	☒Pennsylvania Kids	☒Virginia	☐Arizona

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Jakafi under the patient's prescription drug benefit.

Description:

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
1. Jakafi is indicated for treatment of intermediate or high-risk myelofibrosis (MF), including primary MF, post-polycythemia vera MF and post-essential thrombocythemia MF in adults.
2. Jakafi is indicated for treatment of polycythemia vera (PV) in adults who have had an inadequate response to or are intolerant of hydroxyurea.
3. Jakafi is indicated for treatment of steroid-refractory acute graft-versus-host disease (aGVHD) in adult and pediatric patients 12 years and older.
4. Jakafi is indicated for treatment of chronic graft-versus-host disease (cGVHD) after failure of one or two lines of systemic therapy in adult and pediatric patients 12 years and older.

B. Compendial Uses
1. Symptomatic lower risk myelofibrosis
2. Accelerated phase or blast phase myelofibrosis/acute myeloid leukemia
3. Polycythemia vera in patients with inadequate response or loss of response to interferon therapy
4. Philadelphia chromosome (Ph-like) B-cell Acute Lymphoblastic Leukemia (ALL)/Lymphoblastic lymphoma (LL)
5. Chronic myelomonocytic leukemia (CMML)-2
6. BCR-ABL negative atypical chronic myeloid leukemia (aCML) / Myelodysplastic/Myeloproliferative Neoplasms (MDS/MPN) with neutrophilia
7. Essential Thrombocythemia
8. Myeloid/lymphoid neoplasms with eosinophilia and JAK2 rearrangement in blast phase or chronic phase
9. CAR T-cell-related toxicities - Cytokine release syndrome (CRS)

All other indications are considered experimental/investigational and not medically necessary.

Applicable Drug List:

Jakafi

Policy/Guideline:

Documentation:
Submission of the following information is necessary to initiate the prior authorization review:

A. For Ph-like B-cell acute lymphoblastic leukemia/lymphoblastic lymphoma (LL), medical record documentation confirming either a cytokine receptor-like factor 2 (CRLF2) mutation or a mutation associated with activation of the Janus kinase/signal transducers and activators of transcription (JAK/STAT) pathway.
B. For myeloid and/or lymphoid neoplasms with eosinophilia: Testing or analysis confirming JAK2 rearrangement

Criteria for Initial Approval:

A. **Myelofibrosis/Acute Myeloid Leukemia**
Authorization of 12 months may be granted for the treatment of myelofibrosis/acute myeloid leukemia.

B. **Polycythemia Vera**
Authorization of 12 months may be granted for the treatment of polycythemia vera in members who have had an inadequate response or intolerance to hydroxyurea or peginterferon alfa-2a.

C. **Acute Graft-versus-Host Disease (aGVHD) or Chronic Graft-versus-Host Disease (cGVHD)**
Authorization of 12 months may be granted for the treatment of graft-vs-host disease when any of the following criteria are met:
1. Member has steroid-refractory acute GVHD
2. Member has chronic GVHD and has failed at least one prior line of systemic therapy

D. **Acute Lymphoblastic Leukemia (ALL)/Lymphoblastic Lymphoma (LL)**
Authorization of 12 months may be granted for the treatment of Ph-like B-cell acute lymphoblastic leukemia/lymphoblastic lymphoma for members with either a cytokine receptor-like factor 2 (CRLF2) mutation or a mutation associated with activation of the Janus kinase/signal transducers and activators of transcription (JAK/STAT) pathway.

E. Chronic Myelomonocytic Leukemia (CMML)-2
Authorization of 12 months may be granted for the treatment of chronic myelomonocytic leukemia (CMML)-2 in combination with a hypomethylating agent.

F. Atypical Chronic Myeloid Leukemia (aCML) / Myelodysplastic/Myeloproliferative Neoplasms (MDS/MPN) with Neutrophilia
Authorization of 12 months may be granted for the treatment of BCR-ABL negative aCML or MDS/MPN with neutrophilia as a single agent or in combination with a hypomethylating agent.

G. Essential Thrombocythemia
Authorization of 12 months may be granted for the treatment of essential thrombocythemia in members who have had an inadequate response or intolerance to hydroxyurea, peginterferon alfa-2a, or anagrelide.

H. Myeloid/Lymphoid Neoplasms with Eosinophilia
Authorization of 12 months may be granted for the treatment of myeloid and/or lymphoid neoplasms with eosinophilia and JAK2 rearrangement in the chronic phase or blast phase.

I. Cytokine Release Syndrome
Authorization of 1 month may be granted for treatment of chimeric antigen receptor (CAR) T-cell-induced cytokine release syndrome that is refractory to high-dose corticosteroids and anti-IL-6 therapy.

Continuation of Therapy:

A. Myelofibrosis/Acute Myeloid Leukemia, Polycythemia Vera, Acute GVHD, Chronic GVHD, and Essential Thrombocythemia
Authorization of 12 months may be granted for continued treatment in members requesting reauthorization who have improvement in symptoms and no unacceptable toxicity.

B. Acute Lymphoblastic Leukemia (ALL)/Lymphoblastic Lymphoma (LL), Atypical Chronic Myeloid Leukemia (aCML) / Myelodysplastic/Myeloproliferative Neoplasms (MDS/MPN) with Neutrophilia, Chronic Myelomonocytic Leukemia (CMML)-2, and Myeloid/Lymphoid Neoplasms with Eosinophilia

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization when there is no evidence of unacceptable toxicity or disease progression while on the current regimen.

C. Cytokine Release Syndrome

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

Approval Duration and Quantity Restrictions:

Approval: 12 months

Quantity Level Limit: Jakafi – 60 tablets per 30 days

References:

1. Jakafi [package insert]. Wilmington, DE: Incyte Corporation; December 2022.
2. The NCCN Drugs & Biologics Compendium® © 2023 National Comprehensive Cancer Network, Inc. Available at: http://www.nccn.org. Accessed January 10, 2023.
3. Zeiser R, Burchert A, Lengerke C, et al: Ruxolitinib in corticosteroid-refractory graft-versus-host disease after allogeneic stem cell transplantation: a multicenter survey. Leukemia 2015; 29(10):2062-2068.
4. Zeiser R, Burchert A, Lengerke C, et al: Long-term follow-up of patients with corticosteroid-refractory graft-versus-host disease treated with ruxolitinib. Blood 2016; 128(22):4561
5. Raetz Elizabeth, Loh Mignon. A Phase 2 Study of the JAK1/JAK2 Inhibitor Ruxolitinib with Chemotherapy in Children with De Novo High-Risk CRLF2-Rearranged and/or JAK Pathway-Mutant Acute Lymphoblastic Leukemia. *American Society of Hematology*. 2016: 13(3).
6. Ding YY, Stern JW, Jubelirer TF, et al. Clinical efficacy of Ruxolitinib and chemotherapy in a child with Philadelphia chromosome-like acute lymphoblastic leukemia with GOLGAS-JAK2 fusion and induction failure. *Haematologica*. 2018 Sep;103(9):e427-e431. doi: 10.3324/haematol.2018.192088. Epub 2018 May 17.
7. National Comprehensive Cancer Network. NCCN Clinical Practice Guidelines in Oncology: Pediatric Acute Lymphoblastic Leukemia. Available at: http://www.nccn.org. Version 1.2023. https://www.nccn.org/professionals/physician_gls/pdf/ped_all.pdf. Accessed January 10, 2023.