Jakafi

Description
Jakafi (ruxolitinib)

Background
Jakafi is a Janus Kinase 1 and 2 inhibitor approved by the FDA for the treatment of intermediate or high-risk myelofibrosis, including primary myelofibrosis, post-polycythemia vera myelofibrosis, and post-essential thrombocytopenia myelofibrosis (1).

Myelofibrosis is a disease in which the bone marrow is replaced by scar tissue resulting in blood cells being made in organs such as the liver and the spleen. This disease is marked by an enlarged spleen, anemia, decreased white blood cells and platelets, and myelofibrosis-related symptoms (2).

Polycythemia vera occurs when too many red blood cells are made in the bone marrow. Patients may also experience an increase in white blood cells and platelets. An overabundance of blood cells can cause the spleen to swell, bleeding problems and blood clots in the veins near the skin surface (phlebitis). In addition, it puts patients at increased risk of stroke or heart attack. Jakafi works by inhibiting enzymes called Janus Associated Kinase (JAK) 1 and 2 that are involved in regulating blood and immunological functioning (2).

Regulatory Status
FDA-approved indication: Jakafi is a kinase inhibitor indicated for treatment of: (1)
- Intermediate or high-risk myelofibrosis, including primary myelofibrosis, post-polycythemia vera myelofibrosis and post-essential thrombocytopenia myelofibrosis in adults.
- Polycythemia vera in adults who have had an inadequate response to or are intolerant of hydroxyurea.
• Steroid-refractory acute graft-versus-host disease (GVHD) in adult and pediatric patients 12 years and older.

Jakafi must be held if the patient is experiencing thrombocytopenia with a platelet count below 50x10^9 per L and should not be prescribed in these patients. If the platelet count is below 125 x 10^9 per L dose adjustment may be necessary depending on the current dose of the patient’s medication. CBC monitoring is required every 2-4 weeks until dose is stabilized. CBC w/ diff, palpable spleen length or spleen volume by MRI or CT should be done to monitor disease progression. Patients should be assessed for signs and symptoms of infection. Serious infections should have resolved before starting therapy. Lipid elevations have been reported and lipid levels should be assessed 8-12 weeks from start of therapy and treated as needed (1).

The safety and effectiveness of Jakafi for the treatment of myelofibrosis or polycythemia vera in pediatric patients have not been established. The safety and effectiveness of Jakafi for the treatment of steroid-refractory acute GVHD have been established for the treatment of children 12 years and older (1).

Related policies
Inrebic
Policy
This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Jakafi may be considered medically necessary in patients age 18 years of age or older with intermediate-risk or high-risk myelofibrosis, including primary myelofibrosis, post-polycythemia vera myelofibrosis or post-essential thrombocythemia myelofibrosis, or graft-versus-host disease (GVHD) in allogeneic hematopoietic stem cell transplantation (allo-HCT) and if the conditions indicated below are met.

Jakafi is considered investigational in patients less than 18 years of age and for all other indications.

Prior-Approval Requirements

Age 18 years of age and older

Diagnoses

Patient must have ONE of the following:
1. Intermediate-risk or high-risk myelofibrosis
2. Primary myelofibrosis
3. Post-polycythemia vera myelofibrosis
4. Post-essential thrombocythemia myelofibrosis
5. Polycythemia vera
   a. Inadequate treatment response to or are intolerant of hydroxyurea

**AND ALL** of the following:
   a. Prescriber agrees to assess lipid levels 8-12 weeks from start of therapy and treat it as needed
   b. **NO** serious infections

**Age**
12 years of age and older

**Diagnoses**

Patient must have the following:

1. Acute graft-versus-host disease (GVHD) in allogeneic hematopoietic stem cell transplantation (allo-HCT)
   a. Inadequate treatment response or intolerance to corticosteroid therapy

**AND ALL** of the following:
   a. Prescriber agrees to assess lipid levels 8-12 weeks from start of therapy and treat it as needed
   b. **NO** serious infections

**Prior – Approval Renewal Requirements**

**Age**
18 years of age and older

**Diagnoses**

Patient must have **ONE** of the following:

1. Intermediate-risk or high-risk myelofibrosis
2. Primary myelofibrosis
3. Post-polycythemia vera myelofibrosis
4. Post-essential thrombocythemia myelofibrosis
5. Polycythemia vera
AND the following:
  a. A reduction in palpable spleen length, spleen volume and/or symptomatic improvement

Age 12 years of age and older

Diagnoses

Patient must have the following:

1. Graft-versus-host disease (GVHD) in allogeneic hematopoietic stem cell transplantation (allo-HCT)

AND the following:
  a. Symptomatic improvement

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 6 months

Prior – Approval Renewal Limits

Same as above

Rationale

Summary

Jakafi is a kinase inhibitor indicated for treatment of patients with intermediate or high-risk myelofibrosis, including primary myelofibrosis, post-polycythemia vera myelofibrosis, post-essential thrombocythemia myelofibrosis, and polycythemia vera in patients who have had an inadequate response to or are intolerant of hydroxyurea. Thrombocytopenia, anemia and neutropenia can occur and can be managed by dose reduction, or interruption or transfusion (1).

Prior authorization is required to ensure the safe, clinically appropriate and cost-effective use of Jakafi while maintaining optimal therapeutic outcomes.
References

Policy History

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<td>April 2012</td>
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<td>March 2013</td>
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<td>December 2014</td>
<td>Addition of new indication polycythemia vera who have had an inadequate response to or are intolerant of hydroxyurea</td>
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Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on December 6, 2019 and is effective on January 1, 2020.