Commercial/Healthcare Exchange PA Criteria

Effective: May 12, 2022

Prior Authorization: Pyrukynd

Products Affected: Pyrukynd (mitapivat) oral tablets

Medication Description: Mitapivat is a pyruvate kinase activator that allosterically binds to the pyruvate kinase tetramer and increases pyruvate kinase (PK) activity. The RBC form of pyruvate kinase is mutated in PK deficiency, which leads to reduced adenosine triphosphate, shortened RBC lifespan, and chronic hemolysis

Covered Uses: The treatment of hemolytic anemia in adults with pyruvate kinase (PK) deficiency

Exclusion Criteria: None

Required Medical Information:

1. Diagnosis
2. Medical History

Prescriber Restriction: Prescribed by, or in consultation with, a hematologist

Age Restriction: 18 years of age and older

Coverage Duration: 12 months

Other Criteria:

Initial Approval Criteria

1. **Hemolytic Anemia Due to Pyruvate Kinase Deficiency** – Approve if the patient meets all of the following (A **AND** B)
   - **A.** Patient meets both of the following (i **AND** ii)
     - **i.** Presence of at least two variant/mutant alleles in the pyruvate kinase liver and red blood cell (*PKLR*) gene; **AND**
     - **ii.** At least one of the variant/mutant alleles was a missense variant; **AND**
   - **B.** Patient meets **ONE** of the following (i **OR** ii)
     - **i.** Patient has a current hemoglobin level ≤ 10g/dL; **OR**
     - **ii.** Patient is currently receiving red blood cell transfusions regularly, defined as at least six transfusions within the last year

Renewal Criteria

1. **Hemolytic Anemia due to Pyruvate Kinase deficiency**- Patient is currently receiving Pyrukynd. Approve if the patient meets all of the following (A, B, **AND** C)
   - A. Patient meets both of the following (i **AND** ii)
     - **i.** Prescence of at least two variant/mutant alleles in the pyruvate kinase liver and red blood cell (*PKLR*) gene; **AND**

ii. At least one of the variant/mutant alleles was a missense variant; **AND**

B. Patient has a current hemoglobin level < 13g/dL; **AND**

C. According to the prescriber, the patient has experienced a benefit from therapy based on all of the following (i, ii, **AND** iii)

i. Increase in or maintenance of hemoglobin levels; **AND**

ii. Improvement in or maintenance of hemolysis laboratory parameters; **AND**
Note: Some examples of laboratory parameters that are markers of hemolysis are indirect bilirubin, lactate dehydrogenase, and haptoglobin.

iii. Decrease in or maintenance of transfusion requirements

References:

1. Pyrukynd™ [package insert]. Cambridge, MA, Agios. Updated February 2022. Accessed April 1st, 2022.

Policy Revision history

Rev #	Type of Change	Summary of Change	Sections Affected	Date
1	New Policy	New Policy	All	5/12/2022