JOHNS HOPKINS MEDICINE JOHNS HOPKINS HEALTHCARE	Johns Hopkins HealthCare LLC **Pharmacy Public** **Pharmacy Management Drug Policies**	*Policy Number*	MEDS154
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This document applies to the following Participating Organizations:

Priority Partners

Keywords: Pyrukynd

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I. POLICY

A. Pyrukynd (mitapivat) will require prior authorization for appropriate use. The process for initiating a prior authorization request can be found in policy PHARM 20.
   1. PPMCO members are subject to the Priority Partners formulary, available at www.ppmco.org.
   2. USFHP members are subject to prior authorization criteria, step-edits and days-supply limits outlined in the Tricare Policy Manual. Tricare Policy supersedes JHHC Medical/Pharmacy Policies. Tricare limits may be accessed at: http://pec.ha.osd.mil/formulary_search.php?submenuheader=1

II. POLICY CRITERIA

A. **Pyrukynd** may be approved for patients meeting the following:
   1. Patient is 18 years of age or older
   2. Documentation has been submitted showing the following:
      1. Diagnosis of hemolytic anemia due to pyruvate kinase deficiency
      2. Laboratory findings confirm the presence of at least 2 variant alleles in the pyruvate kinase liver and red blood cell (PKLR) gene, with at least 1 allele being a missense variant
      3. Patient is not homozygous for the R479H mutation or have 2 non-missense mutations, without the presence of another missense mutation in the PKLR gene
      4. One of the following:
         1. Patient has a current hemoglobin level ≤10g/dL
         2. Patient is currently receiving red blood cell transfusions regularly, defined as at least 6 transfusions within the last year
      5. Patient will be on a concurrent regimen with oral folic acid
      6. Prescriber is, or has consulted with, a hematologist

III. AUTHORIZATION PERIOD/LIMITATIONS

A. Initial approval will be limited to 6 months of therapy
B. Approval for continuation of therapy may be extended in 12-month intervals with documentation of the following:
   1. Patient has had a beneficial response to treatment, evidenced by one of the following:

1. Hemoglobin increase of ≥ 1.5 g/dL from baseline
2. Decrease in the number of RBC transfusions from baseline

2. Patient will continue to take a concurrent regimen with oral folic acid

C. Limitation:
1. Pyrukynd will not be approved for continuation if the patient has not experienced a clinical benefit by 24 weeks of therapy.
2. If continuation is not approved: A one-time approval of the appropriate dose taper pack will be issued to support gradual discontinuation of Pyrukynd, and reduce the risk of acute hemolysis

IV. EXCLUSIONS

A. Pyrukynd will not be approved for the following:
1. Pediatric patients
2. Patients with moderate or severe hepatic impairment
3. Patients who have had a prior bone marrow or stem cell transplant
4. Patients with a history of splenectomy, or planning to undergo splenectomy
5. Patients taking strong CYP3A inhibitors or inducers
6. Any indications or usage that is not FDA-approved, or guideline-supported

B. The use of physician samples, or manufacturer product discounts, does not guarantee coverage under the provisions of the medical and/or pharmacy benefit. All pertinent criteria must be met in order to be eligible for benefit coverage.

V. REFERENCES

1. Pyrukynd [prescribing information]. Cambridge, MA: Agios Pharmaceuticals Inc; February 2022.

VI. APPROVALS

Signature on file at JHHC

DATE OF REVISION	SUMMARY OF CHANGE
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