

AvMed

PHARMACY PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request. All other information may be filled in by office staff; **fax to 1-305-671-0200**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If the information provided is not complete, correct, or legible, the authorization process can be delayed**

Drug Requested: Kygevvi® (doxecitine and doxribtimine)

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member AvMed #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Name/Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy:** _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight (if applicable): _____ **Date weight obtained:** _____

Recommended Dosing: Kygevvi dosing is weight-based and includes three recommended dosage levels. Titrate to the next dosage level based on tolerability after a minimum of 2 weeks at the current dose. Administer Kygevvi orally in 3 equally divided doses with food.

Kygevvi Dosage Level	Kygevvi Dosage (mg/kg/day)
Starting	260 mg/kg/day (consisting of 130 mg doxecitine and 130 mg doxribtimine)
Intermediate	520 mg/kg/day (consisting of 260 mg doxecitine and 260 mg doxribtimine)
Maintenance	800 mg/kg/day (consisting of 400 mg doxecitine and 400 mg doxribtimine)

Quantity Limit: 16 packets per day

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Number of Packets Based on Total Daily Dose per Prescribing Information

Total Daily Dose (mg/day)	Total Number of Kygevi Packets for Reconstitution
750 – 4,049	1
4,050 – 8,249	2
8,250 – 12,749	3
12,750 – 17,249	4
17,250 – 21,749	5
21,750 – 24,749	6
24,750 – 29,249	7
29,250 – 33,749	8
33,750 – 38,249	9
38,250 – 42,749	10
42,750 – 47,249	11
47,250 – 51,749	12
51,750 – 54,749	13
54,750 – 59,249	14
59,250 – 63,749	15
63,750 – 68,249	16

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Authorization: 12 months

- ☐ Medication must be prescribed by or in consultation with a neurologist, geneticist, pulmonologist or specialist experienced in managing mitochondrial myopathies
- ☐ Member has a diagnosis of thymidine kinase 2 deficiency (TK2d) confirmed by genetic testing of pathogenic TK2 variants (**submit documentation**)
- ☐ Member has clinical documentation of initial symptom onset at ≤ 12 years of age, consistent with infantile- or childhood-onset TK2d (**submit documentation**) [**NOTE: Examples of symptoms consistent with TK2d include progressive muscle weakness, hypotonia (i.e., low muscle tone), respiratory insufficiency, loss of motor skills, feeding and/or swallowing difficulties, facial weakness/paralysis, ptosis (i.e., drooping eyelid[s]), ophthalmoparesis (i.e., difficulty moving the eyes), developmental delay and regression, hearing loss, and seizures**]

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- ☐ Member has an absence of other genetic disease, polygenic disease, or inborn errors of metabolism
- ☐ Member has baseline liver transaminases (ALT, AST) and total bilirubin levels prior to treatment initiation (**submit documentation**)
- ☐ Member does **NOT** have a history of liver disease or liver function tests (ALT, AST, or total bilirubin) greater than or equal two times the upper limit of normal at baseline per the laboratory administering the test
- ☐ Member does **NOT** have renal insufficiency requiring dialysis
- ☐ Member does **NOT** have severe end-organ hypo-perfusion syndrome secondary to cardiac failure resulting in lactic acidosis

Reauthorization: 12 months. Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

- ☐ Member has demonstrated clinical benefit to therapy as evidenced by improvement or stabilization in motor function (e.g., 6-minute walk test [6MWT], Motor Function Measure [MFM] 20 or MFM 30), pulmonary function or ability to meet growth and developmental milestones) (**submit documentation**)
- ☐ Member has **NOT** experienced any unacceptable toxicity from the drug (e.g., diarrhea and vomiting leading to hospitalization, hepatic toxicity, elevated liver enzymes)

Medication being provided by Specialty Pharmacy – Proprium Rx

Not all drugs may be covered under every Plan

If a drug is non-formulary on a Plan, documentation of medical necessity will be required.

*****Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.*****

****Previous therapies will be verified through pharmacy paid claims or submitted chart notes.****