RX.PA.041.MPC *Nulibry® (fosdenopterin)*

PURPOSE

Nulibry® is a cyclic pyranopterin monophosphate (cPMP) indication to reduce the risk of mortality in patients with molybdenum cofactor deficiency (McCD) Type A. Maryland Physicians Care requires Prior Authorization for its use.

Molybdenum Cofactor Deficiency Type A - Approve Nulibry if the member meets **ALL** of the following conditions:

A. Initial Therapy:

a. Must be prescribed by, or in consultation with, a neonatologist, a pediatric neurologist or specialist with expertise in the treatment of inherited metabolic disorders
AND

b. Clinical documentation for the diagnosis of molybdenum cofactor deficiency type A:
   i. Genetic testing confirmation of a mutation in the MOCS1 gene
AND

c. Documentation of clinical and/or biochemical features associated with MoCD Type A
   i. Seizures
   ii. Limb/axial hypertonia
   iii. Low serum uric acid
   Normal range:
      - Adult male: 4.0-8.5 mg/dL or 0.24-0.51 mmol/L
      - Adult female: 2.7-7.3 mg/dL or 0.16-0.43 mmol/L
      - Elderly: A slight increase in values may occur
      - Child: 2.5-5.5 mg/dL or 0.12-0.32 mmol/L
      - Newborn: 2.0-6.2 mg/dL
   iv. Elevated urinary xanthine and hypoxanthine
   Normal range:
      - Xanthine: < 40mol/L
      - Hypoxanthine: < 70 mol/L
   v. Elevated sulfites in urine
   Normal range:
      - 7-47 mmol/24 hours

d. Dose does not exceed 0.9mg/kg once daily

B. Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling.

C. Nulibry will be considered investigational or experimental for any other use and will not be covered.

D. <u>Continuation of therapy:</u>

- MPC Renewal:
  a) Member has a documented clinical response to therapy as stabilization or improvement as determined by the prescriber.
  b) Prescribed by, or in consultation with, a neonatologist, a pediatric neurologist or specialist with expertise in the treatment of inherited metabolic disorders.
- Renewal from Previous Insurer:
  a) Members who have received prior approval (from insurer other than MPC), or have been receiving medication samples, should be considered under criterion A (Initial Therapy)
  b) Provider has a documented clinical response of the member's condition which has stabilized or improved compared to baseline.

<u>Approval Duration:</u>

A. Initial Therapy: Approve for 6 months
B. Continuation of Therapy: Approve for 1 year

CPT Code	Description
J3490	Unclassifed drugs Single-dose vial of 9.5mg

References:

1. Nulibry® injection [prescribing information]. Boston, MA: Origin Biosciences, Inc.; February 2021.

REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Annual Review *Change in Non-MPC renewal to renewal from previous insurer*	*02/2024*
Annual review	*02/2023*
Selected Revision *Addition of MPC vs Non-MPC Renewal Criteria*	*09/2022*
Annual review	*02/2022*
Addition of dosing requirements and off-label restrictions	*12/2021*
P&T Review	*11/2020*