PHARMACY POLICY STATEMENT
Marketplace

DRUG NAME	Palynziq (pegvaliase-pqpz)
BILLING CODE	Must use valid NDC
BENEFIT TYPE	Pharmacy
	Prior Authorization Required

Palynziq, approved by the FDA in 2018, is a phenylalanine (Phe)-metabolizing enzyme indicated to reduce blood Phe concentrations in adult patients with phenylketonuria (PKU) who have uncontrolled blood Phe concentrations greater than 600 micromol/L on existing management. Palynziq is only available through a REMS program due to a risk of anaphylaxis.
PKU results from a deficiency of phenylalanine hydroxylase (PAH) enzyme, leading to increased concentrations of Phe. If untreated, this excess accumulation causes neuropsychiatric and neurocognitive symptoms. Palynziq is a PEGylated phenylalanine ammonia lyase (PAL) enzyme that converts phenylalanine to ammonia and trans-cinnamic acid. It works as an enzyme substitution therapy as PAL substitutes for the deficient PAH enzyme activity. Standard of care for PKU is a Phe-restricted diet.

Palynziq (pegvaliase-pqpz) will be considered for coverage when the following criteria are met:

Phenylketonuria (PKU)

For **initial** authorization:

1. Member is at least 18 years of age; AND
2. Medication must be prescribed by or in consultation with specialist experienced in metabolic or genetic diseases; AND
3. Member has a diagnosis of phenylketonuria; AND
4. Member has uncontrolled blood phenylalanine (Phe) concentrations greater than 600 micromol/L on existing management with Kuvan* (requires prior authorization) in conjunction with following recommended dietary modifications; AND
5. Palynziq will not be prescribed in combination with Kuvan.
6. **Dosage allowed/Quantity limit:** Initial, 2.5 mg subQ once weekly x 4 weeks. Titrate over at least 5 weeks to 20 mg once daily. May increase to 40 mg daily after 24 weeks on 20 mg/day if control not achieved. May increase to 60 mg daily if control not achieved with 40 mg/day after 16 weeks. Discontinue after 16 weeks of 60 mg/day if adequate response not achieved. (Max dose 60 mg/day). QL: 90 syringes per 30 days

**Note*: *A trial of Kuvan is not necessary if there is documentation of 2 null mutations. However, a trial and failure of compliant diet management is still required.*

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must show at least one of the following:
   a) Member has achieved at least a 20% reduction in blood phenylalanine concentration from pretreatment baseline;
   b) Member has achieved a blood phenylalanine concentration of 600 micromol/L or less.

If all the above requirements are met, the medication will be approved for an additional 12 months.