



Drug Name: Palynziq (pegvaliase-pqpz)

Effective Date: 6/7/19

Review Date: 6/2019, 6/2020

Drug Name: Palynziq (pegvaliase-pqpz)	
Required Medical Information:	<p>Initial</p> <ul style="list-style-type: none"> • Patient is ≥ 18 years of age • Patient has a diagnosis of phenylketonuria (PKU) • Prescriber must be certified with Palynziq REMS program • Patient currently uncontrolled on existing management and has a blood phenylalanine concentrations greater than 600 micromol/L (labs to be provided) • Patient has failed therapy with Kuvan • Patient must have prescription of auto-injectable epinephrine on hand at all times • Patient has failed therapy with dietary restriction of phenylalanine • Palynziq is not being used in combination with Kuvan (sapropterin) <p>Renewal – initial (on maintenance dose of 20mg daily for less than 3 months)</p> <ul style="list-style-type: none"> • Patient has been titrated to 20mg daily, or lowest effective and tolerated dose • Blood phenylalanine concentration after treatment initiation (labs to be provided) • Palynziq is not being used in combination with Kuvan (sapropterin) <p>Renewal – maintenance and/or 40mg daily dose</p> <ul style="list-style-type: none"> • Palynziq is not being used in combination with Kuvan (sapropterin); AND • Blood phenylalanine concentration lab results provided; AND • If requesting new daily dose of 40mg: Documentation that member has been adherent to 20mg daily and requires increase to 40mg once daily because $<20\%$ reduction in blood phenylalanine concentration from baseline or blood phenylalanine concentration >600 micromol/L; OR • If requesting maintenance daily dose of 20 mg or 40mg: $\geq 20\%$ reduction in blood phenylalanine concentration from baseline OR blood phenylalanine concentration less than or equal to 600 micromol/L



Quantity Limit/Coverage duration:	<p>Initial: 3 months</p> <p>Renewal:</p> <ul style="list-style-type: none">• Dose of 20mg once daily: 6 months, 30 ml per 25 days• Dose of 40mg once daily (initial): 4 months, 60 ml per 25 days• Dose of 40mg once daily (maintenance): 6 months, 60 ml per 25 days