Pharmacy Drug Policy & Procedure

Policy Name:	Kuvan, sapropterin pack and tablets	Policy #:	1533P

Purpose of the Policy

The purpose of this policy is to establish the criteria for coverage of Kuvan.

Statement of the Policy

Health Alliance Medical Plans and Health Alliance Northwest will approve the use of Kuvan under the Specialty Pharmacy benefit when the following criteria have been met.

Criteria

1. Coverage Criteria for Hyperphenylalaninemia (HPA) due to tetrahydrobiopterin-responsive Phenylketonuria (PKU)

1.1 Documented diagnosis of Phenylketonuria (PKU)
1.2 Treated by a specialist knowledgeable in the management of PKU
1.3 Documentation that therapy will accompany a strict Phe-restrictive diet
1.4 Baseline Phe level:
- >6mg/dL (360 micromol/L) if 12 years of age, OR
- >10 mg/dL (600 micromol/L) if >12 years of age

1.5 For brand Kuvan, documented trial and failure with generic sapropterin tablets/packets

2. Exclusion Criteria

2.1 Documented non-response to Kuvan indicated by a failure to reduce baseline Phe level by 30%.
2.2 Kuvan will not be approved if the member is also receiving Palynziq because there is no data available to support the use of concomitant therapy with these medications in the treatment of PKU

3. Approval Time

3.1 Initial approval
- 12 months with a starting dose of 10mg/kg/day to 20mg/kg/day

3.2 Continued approval
- 12 months if the following is met:
  - At least a 30% reduction in the baseline level following 1 continuous month of therapy
  - Documented compliance with Kuvan
  - Documented compliance with a Phe-restricted diet
  - Member is followed by a specialist knowledgeable in the management of PKU
- Note: Maintenance dose range from 5–20 mg/kg/day

CPT Codes

HCPCS Codes

References

1. Kuvan (sapropterin) [prescribing information]. Novato, CA: BioMarin Pharmaceutical Inc; February 2021.
2. Keil S, Anjema K, van Spronsen FJ, et al. Long-term follow-up and outcome of phenylketonuria patients on sapropterin: a retrospective study. Pediatrics 2013; 131:e1881.
3. Levy HL, Milanowski A, Chakrapani A et al. Efficacy of sapropterin dihydrochloride (tetrahydrobiopterin, 6R-BH4) for reduction of phenylalanine concentration in patients with phenylketonuria: a phase III randomised placebo-controlled study. Lancet. 2007; 370(9586):504–510.
4. Vockley J, Andersson HA, Antshel KM, et al. Phenylalanine hydroxylase deficiency: diagnosis and management American College of Medical Genetics and Genomics (ACMG) Guideline. Genet Med. 2014 Feb;16(2):188-200.

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