



HSE Prescribing Protocol

Sapropterin Dihydrochloride (Kuvan®)

for

**Treatment of Hyperphenylalaninaemia in Adults and Paediatric Patients with
Phenylketonuria and Tetrahydrobiopterin Disorders**

This document is intended for use by healthcare professionals only.

This guideline should be used in conjunction with the full prescribing and administration details in the sapropterin (Kuvan®) Summary of Product Characteristics (SmPC)

https://www.ema.europa.eu/en/documents/product-information/kuvan-epar-product-information_en.pdf

INDICATION FOR USE¹

TREATMENT	HSE APPROVED INDICATION	ICD10	PROTOCOL CODE
Sapropterin dihydrochloride (Kuvan®)	For the treatment of hyperphenylalaninaemia (HPA) in adults and paediatric patients of all ages with phenylketonuria (PKU) and tetrahydrobiopterin (BH4) deficiency who have been shown to be responsive to such treatment.	E70	ERT003

TREATMENT¹

TREATMENT	DOSE	ROUTE	FREQUENCY	RESPONSE
Sapropterin (Kuvan®) 100mg soluble tablets				
<p>Phase 1 Loading test</p> <p>The 48-hour loading test is carried out in an inpatient or outpatient setting (at the discretion of the physician).</p>	<p>The loading test requires a sapropterin dose of 20mg/kg administered once daily for a total of two doses only.</p> <p>The second dose is administered 24 hours after the first loading dose.</p> <p>Blood Phe samples are taken at 0, 8, 16 and 24 hours after each sapropterin administration. Correct blood sampling is crucial for a meaningful interpretation of the test.</p> <p>The calculated daily dose based on body weight should <u>always</u> be rounded to the nearest multiple of 100mg.</p>	PO	<p>Once daily (Single daily dose at the same time each day preferably in the morning)</p>	<p>A satisfactory response is defined as a reduction in the blood Phe concentration of at least 30% from baseline.</p> <p>If there is less than a 30% response, longer test durations can be considered (max 7 days) for those with suggestive phenotypes and/or genotypes.</p> <p>Only patients with a satisfactory response to Phase 1 can progress to Phase 2.</p>

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Protocol Code: ERT003	Approved by: Dr Mike O'Connor National Clinical Advisor & Group Lead, Acute Hospitals	Contributors: HSE National ERT Steering Committee Adult Metabolic Services: J Ivory, C Newman-Thacker, B Gillman Paediatric Metabolic Services: Dr K Bhattacharya, O Walsh, J McNulty, M Irranca,	Page 2 of 10

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	DOSE	ROUTE	FREQUENCY	RESPONSE
Phase 2 Sapropterin Trial Optimisation of dose	Patients are started on a dose of sapropterin 10mg/kg/day (rounded to the nearest 100mg) initially which can be titrated weekly, to a dose between 1 to 20mg/kg/day. This should be completed within two months of initiation of phase 2. Patients are required to submit weekly blood Phe samples during phase 2.	PO	Once daily	Satisfactory response / optimum dosing in Phase 2 is defined as a dose which leads to the achievement of greater than or equal to 75% of blood Phe levels within the target range of Phe less than 600 µmol/L for patients 12 years or older, and Phe less than 360 µmol/L for patients less than 12 years, and as a ≥ 100 % increase in natural protein intake. Only patients with a satisfactory response to Phase 2 can progress to Phase 3.
Phase 3 Long-Term Six Month Sapropterin Trial	Patients are required to submit weekly blood Phe samples during phase 3. The dose of sapropterin can be adjusted (1-20mg/kg/day) to achieve the defined response.	PO	Once daily	Long-term sapropterin responsiveness is defined as greater than or equal to 75% of blood Phe levels within the target range (Phe less than 600 µmol/L for patient 12 years of age or older, and less than 360 µmol/L for patients under 12 years of age) and a ≥ 100 % increase in natural protein intake. Patients who have successfully completed the long-term six-month sapropterin treatment trial and are deemed to be responders and are considered eligible for continued long-term treatment.

For those >18 years dose should be capped at a weight equivalent to a body mass index of 27kg/m².

Treatment must be initiated and supervised by a physician experienced in the treatment of PKU and BH4 deficiency. Active management of dietary phenylalanine and overall protein intake while taking

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this medicinal product is required to ensure adequate control of blood phenylalanine levels and nutritional balance.¹

ELIGIBILITY CRITERIA

- Indication as above

To be eligible for treatment with sapropterin patients must:

- Successfully complete a three-phase initiation protocol to determine short and long-term responsiveness to sapropterin.
- Meet all of the starting criteria outlined below for each phase of the protocol.
- Have confirmation of variants of the PAH gene (this confirmation is not required for neonates).

Starting Criteria Phase 1 Sapropterin Loading Test

- A diagnosis of PKU which requires dietary treatment to keep Phe levels within age-appropriate target range
- The natural protein intake is maximised prior to conducting the loading test
- Complete baseline assessments of blood Phe concentrations - 3 or more blood Phe levels must be obtained within the month prior to the start of the loading test
- Patient/carer has received appropriate counselling regarding sapropterin therapy and diet
- Discussion with patients/caregivers regarding the expected outcomes of therapy including the possibility of treatment discontinuation and signed consent

Note: In the **neonatal period** a loading test can be done before starting the diet but should not be longer than 24 hours to avoid delays in treatment.

Patients with two known sapropterin responsive mutations are not required to undergo a 48-hour loading test and are recommended to directly proceed to the Phase 2 of the protocol.

Starting Criteria Phase 2 Sapropterin Trial Optimisation of dose

- Demonstrated sapropterin responsiveness as defined in the TREATMENT table above
- Patient/carer agrees to send weekly blood Phe levels
- Patient/carer agrees to complete a nutritional assessment each week which is used to estimate nutrient/Phe intake and assess nutritional status

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Starting Criteria Phase 3 Long-Term Six Month Sapropterin Trial

- Demonstrated sapropterin responsiveness as defined in the TREATMENT table above
- Patient/carer agrees to complete a nutritional assessment each month which is used to estimate nutrient/Phe intake and assess nutritional status

EXCLUSION CRITERIA

Patients who do not satisfy any of the above criteria or who meet any of the exclusionary criteria below are not considered eligible for the loading test or treatment with sapropterin.

- Patients with a diagnosis of PKU which does not require any dietary treatment including synthetic protein supplementation
- Two known null mutations in the PAH gene
- Patients with untreated Phe levels less than 360µmol/L (or 600µmol/L if aged 12 or over)
- Patient/carer declines a loading test when indicated
- The patient is unable to comply with the associated monitoring criteria, including attending all required clinic visits

CONTRAINDICATIONS

- Hypersensitivity to sapropterin dihydrochloride or to any of the excipients

BASELINE TESTS AND MONITORING

Complete baseline assessments of blood Phe concentrations - 3 or more blood Phe levels must be obtained within the month prior to the start of the loading test.

Monitoring Requirements during Phase 3 Long-Term Six Month Sapropterin Treatment Trial Period

Minimum Monitoring Requirements and Follow-Up of Patients during Phase 3

Monitoring Parameters	Frequency of Monitoring	Requirements	Monitored by	Notes
Blood Phe levels	Weekly	≥75% must be within target range	Metabolic team	If inadequate control of blood Phe levels is observed, review adherence to prescribed treatment, and diet, before considering a dose adjustment. ¹

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Natural protein intake	Monthly by nutritional assessment	Increased as appropriate	Dietitian	
Synthetic protein supplements	Monthly	Decreased as clinically indicated	Dietitian & clinician	
Sapropterin dose review	3 monthly	Titrated to lowest dose necessary	Clinician	The sapropterin dose is titrated to the lowest dose necessary to maintain a response. Blood Phe levels must be maintained within the target range with sapropterin treatment and diet relaxation. ^{1,4,6}
Nutritional bloods	As clinically required	FBC, B12, Folate, Ferritin, Vit D, plasma amino acids	Metabolic team	Monitoring dietary patterns, micronutrient intake, and biochemical nutritional status should be performed as required, particularly in patients who have stopped or reduced intake of protein substitute.

Monitoring Requirements during Continued Long-Term Treatment with Sapropterin

- Review the on-going prescription for sapropterin as appropriate at each clinic visit – at least 6-12 monthly.⁴
- Monitor the need for vitamin and mineral supplements at each clinic visit.
- Sapropterin therapy still requires adherence to an agreed dietary regimen and regular monitoring of blood Phe levels. Blood Phe levels are monitored regularly as per age-appropriate local guidelines during the first year of long-term treatment.⁴

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SPECIAL WARNINGS AND PRECAUTION FOR USE

See SmPC for full details.

STOPPING CRITERIA

Patients will cease sapropterin treatment at any time during the 3-Phase protocol period or during continued long-term treatment if any of the following criteria apply:

- Patients are unable to tolerate sapropterin due to severe adverse events. Please consult SmPC for details on adverse effects¹
- Blood Phe levels consistently exceed the upper target range and no improvement associated with any increase in sapropterin dosage during Phase 3
- Nutritional status deteriorates as determined by the physician and dietitian
- Patients fail to attend their metabolic clinic as instructed for assessment and monitoring during the 3-Phase protocol period
- Patients on continued long-term treatment with sapropterin who fail to attend their metabolic clinic for assessment and monitoring as per age appropriate European PKU guidelines

ADVERSE EFFECTS

See SmPC for full details

DRUG INTERACTIONS

See SmPC. Caution is recommended when using methotrexate, trimethoprim, glyceryl trinitrate, isosorbide dinitrate, sodium nitroprusside, molsidomin, phosphodiesterase type 5 inhibitors, minoxidil and levodopa.

ATC CODE

Sapropterin A16AX07

REIMBURSEMENT CATEGORY

Patients within the public health system will be funded for their sapropterin treatment by the Health Service Executive (HSE).

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ADDITIONAL INFORMATION

Phenylketonuria (PKU) is a rare genetic disorder which prevents the breakdown of the amino acid phenylalanine (Phe) causing accumulation in the body. High levels of Phe are extremely toxic to the brain and untreated PKU causes profound brain damage resulting in very low IQ, seizures, muscle stiffness, autism, and persistent behavioural problems. In pregnancies of women with PKU, the foetus can be affected by high levels of Phe. Sapropterin (Kuvan®) treatment aims to lower the blood Phe levels to close to or below the European Guideline levels as outlined in the table below:

Patient Group	Target Blood Phe levels (µmol/L)
Patients with PKU ≤12 years	120–360
Patients with PKU > 12 years	120–600
Patients who are planning a pregnancy/are pregnant	120-360

The treatment outlined in this guideline should be initiated in an appropriate setting for the management of PKU or Tetrahydrobiopterin (BH4) Deficiency. Paediatricians will undertake to refer patients to the adult centre on reaching the age of 16-18 years. Adult and paediatric centres undertake to ensure as much as possible a seamless transfer of care.

Collaboration between the tertiary treatment centres and local primary and secondary care services is imperative to ensuring PKU patients receive high standards of care. Local primary and secondary care clinicians will undertake to ensure all PKU patients are referred to the specialist team in the centre for review/ input. They will endeavour to support local colleagues wherever necessary.

Prior to commencing treatment there should be a full discussion with patients/caregivers regarding the expected outcomes of therapy including the possibility of treatment discontinuation.

Special Considerations

At the discretion of the clinician, in line with international standards, sapropterin therapy may be considered for cases of maternal PKU (if women are known to be sapropterin responders +/- dietary treatment alone has failed), or for cases of hyperphenylalaninaemia in adults and paediatric patients of all ages with BH4 deficiency (e.g. 6-Pyruvoyl tetrahydropterin synthase (PTPS) deficiency) who have been shown to be responsive to such treatment.

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APPENDIX

The sapropterin guidelines have been reviewed by a group of prescribing physicians and healthcare professionals working in the treatment centres of excellence in Ireland. The guidelines are designed to standardise practice and support the implementation of treatment pathways for these patients in Ireland.

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Revision History

Version	Changes
1.0 2018	Superseded
2.0 2022	<p>3.1.3 The 48 hour loading test can now be carried out in an inpatient or outpatient setting as per practical experience.</p> <p>3.2.1 The Phe blood testing requirements have reduced from twice weekly to weekly based on practical experience. The turn-around time for blood tests made clinical interpretation of dose and diet adjustments difficult.</p> <p>3.2.2 The time to complete phase 2 of the protocol has been extended from one month to two months. This is based on practical experience as majority of patients starting on 10mg/kg/day will need longer than 4 weeks to reach an optimum dose.</p> <p>3.2.3 /3.3.3 The definition of clinical response to sapropterin has been amended removing the requirement for a 50% reduction in synthetic protein to “with a significant decrease in synthetic protein requirement clinically appropriate to the nutritional status of the individual patient.” This is in line with European PKU Guidelines.</p> <p>3.3 Phase 3 of the protocol has been extended from 5 months to 6 months based on practical experience to optimise longer-term dose response.</p>
3.0 2025	Layout changed to align with other AIDMP Protocols. Removed table of contents. Added table with indication, ICD10 code and protocol code, table with dose, route, frequency, response. Additional information from SmPC added. Phase 1 Loading test, longer test durations can be considered (max 7 days) for those with suggestive phenotypes and/or genotypes. Phase 2 and 3 responses updated. Removed: an increase in protein exchanges (at least 2-fold) with a significant decrease in synthetic protein requirement clinically appropriate to the nutritional status of the individual patient. Added: ≥ 100 % increase in natural protein intake. Frequency of dose review from monthly to 3 monthly. Nutritional bloods added to monitoring parameters. References updated.

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