PROCEDURE TITLE: Phenylketonuria (New Diagnosis)	Document No: Version No:

Document Type: Clinical	
Procedure Owner: Grainne Bauer **Title: Chief Director of Nursing**	**Procedure Author: Louise Perris** **Title: Clinical Nurse Education Facilitator,** **National Centre for Inherited Metabolic Disorders.**
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1. PROCEDURE STATEMENT

Kuvan (Sapropterin Dihydrochloride) is a prescription medication which is used to lower phenylalanine levels in children with a specific type of Phenylketonuria (PKU).

1.0 PURPOSE

This document provides direction to staff of Children's Health Ireland (CHI) to

1.1 Complete a trial of Sapropterin Dihydrochloride (Kuvan®) to ascertain if the patient is responsive to the drug.

1.2 To stabilise Phenylalanine (Phe) levels.

1.3 To establish special feeding regime if required

1.4 To provide emotional support and education to parents

2.0 SCOPE:

2.1 This procedure applies to CHI healthcare workers who are full time or part time, permanent or temporary, including agency and contractors, and also applies to students attending clinical placements in CHI.

2.2 Target Patient Population: all children and young people attending CHI as inpatients or day cases.

3.0 DEFINITIONS / ABBREVIATIONS:

Phe: Phenylalanine

PKU: Phenylketonuria

Kuvan®: Sapropterin Dihydrochloride

CHI: Children's Health Ireland

OPD: Out Patients Department

5.0 ROLES and RESPONSIBILITIES

It is the responsibility of:

5.1 All healthcare professionals and students to:

Be aware of, read and practice in accordance with this procedure.

5.2 Heads of the department to:

Ensure staff in their area are aware of, have read and understand, and attend any required training, in relation to this procedure.

5.3 Document Owner

Ensure the procedure is communicated and disseminated to all relevant staff.

6.0 PROCEDURES

6.1 Initial Assessment

6.1.1 Assess patient and record baseline PEWS score in age-appropriate PEWS chart on electronic healthcare record. Reassess as clinically indicated. Escalate concerns to senior nurse and medical team as appropriate.

6.1.2 Ensure patient/parent/carer is provided with information about PKU and aware of the plan of care for the hospital admission.

6.1.3 Ensure parent/carer is fully informed about what Sapropterin Dihydrochloride (Kuvan®) is and that the protocol for the Sapropterin Dihydrochloride (Kuvan®) trial is explained. Informed consent to be obtained by the metabolic team from the parents.

6.1.4 Obtain child's weight. Refer to the patient Weighing Policy.

6.1.5 Ensure 20mg/kg of Sapropterin Dihydrochloride (Kuvan®) is prescribed by the medical team.

6.2 During Sapropterin Dihydrochloride (Kuvan®) trial:

6.2.1 Refer to the Management Protocol for Sapropterin Dihydrochloride (Kuvan®) Testing for Neonates. Assist with initial blood work. A confirmatory liquid sample for phenylalanine and tyrosine should be obtained. Ensure lab form is correctly labelled 'For Phenylalanine/Tyrosine'. Ensure Metabolic Investigation Lab form is used. Tubes must be filled completely.

6.2.2 Commence Sapropterin Dihydrochloride (Kuvan®) trial if confirmatory sample phenylalanine level is >360. An acylcarnitine card sample for phenylalanine and tyrosine for time 0 (baseline) should be obtained– one circle is sufficient. A sample to rule out Pterin defect should also be obtained at this time- 5 circles on an acylcarnitine card. Ensure that samples are labelled correctly and transferred to laboratory.

6.2.3 Administer Sapropterin Dihydrochloride (Kuvan®) as prescribed as soon as possible post - time 0 (baseline) bloods.

6.2.4 Continue regular feeding (breast milk or formula). Ensure that fluid balance is recorded accurately for duration of the trial.

6.2.5 Collect samples on acylcarnitine card at time 4, 8, 12, 16 and 24 hours. These samples are timed from the actual time Sapropterin Dihydrochloride (Kuvan®) is ingested. Collect liquid phenylalanine level at time 16 hours post ingestion or as otherwise requested by the metabolic consultant.

6.3 Dietary interventions:

6.3.1 Commence diet as per the direction of metabolic consultant post Sapropterin Dihydrochloride (Kuvan®) trial in conjunction with the metabolic dietitian. Frequently a synthetic feed may be commenced at this time until results from the trial are available.

6.3.4 Refer to daily dietary Flow Sheet on electronic healthcare record for direction in relation to prescribed volume. Check all feeds and volumes with a second staff member prior to administration (one of who must be a registered nurse.

6.3.5 Introduce Natural Protein formula as directed by Consultant and metabolic dietitian. Bottle feeding: Offer natural protein feed first followed by synthetic feed. Breast feeding: Offer synthetic feed followed by breast feed. Record Intake and Output accurately in fluid balance.

6.3.6 Obtain liquid phenylalanine levels daily or as otherwise directed by metabolic team. Take phenylalanine samples via heel prick before morning feed. Note for same day results ensure sample is received in the laboratory by 10am.

6.3.7 Liaise daily with dietitian in relation to the adjustment of feed volume in response to Phenylalanine levels. Refer to daily diet sheet on electronic healthcare records for direction.

6.3.8 Weigh weekly and plot on a weight chart while in hospital to ensure adequate protein for growth and development is being delivered in diet. Plot on weight chart.

6.4 Discharge

6.4.1 Commence Parental Education in liaison with Metabolic Clinical Nurse Specialist and Dietetic Team in relation to the condition, the dietary management and rationale for same, implications of non-compliance, blood sampling and mode of inheritance.

6.4.2 Ensure that parents have received education and training with blood-letting for Phenylalanine (Phe) levels in the parental competency.

6.4.3 Ensure that parents/carers are signed off with medication administration competency if required.

6.4.4 Clarify with dietitian the plan for diet for the patient on discharge and ensure that parents are fully up to date with dietary information.

6.4.5 Liaise with Metabolic team in relation to scheduling of consults to other disciplines and screening of siblings.

6.4.6 Ensure that prescription is completed if patient is responsive to Sapropterin Dihydrochloride (Kuvan®) before discharge. Obtain pharmacy details from parent(s). Ensure that specific Sapropterin Dihydrochloride (Kuvan®) request for product form has been completed by appropriate Consultant and returned to pharmacy in a timely manner. Provide parent/guardian with Sapropterin Dihydrochloride (Kuvan®) information card for local pharmacy on discharge. This card is to be given to local pharmacy if patient is responsive and commencing extended treatment of Sapropterin Dihydrochloride (Kuvan®).

6.4.7 Complete patient discharge and ensure that a follow up OPD appointment as requested by metabolic consultant is communicated to parents.

7.0 COMMUNICATION AND TRAINING:

7.1 Communication and Dissemination: This guideline will be posted on Q-Pulse and the internet, and disseminated to relevant departments.

7.2 Training: Education and training will be delivered at departmental/unit level using existing educational resources, e.g. Clinical Nurse Education Facilitators. Education is included in induction packages for relevant clinical areas/staff.

8.0 COMPLIANCE MONITORING:

8.1 This procedure will be reviewed and updated by the owner or their designee within a three-year cycle, or earlier if required due to updated guidance, evidence or legislation.

9.0 APPROVAL:

Proposed: Fionnuala O Neill, ADON, CHI

Concurred: CHI Nurse Practice Committee

Approved: Grainne Bauer Chief Director of Nursing on behalf of the CHI Nursing Documentation Approval Committee **October 2025**

10.0 VERSION CONTROL HISTORY:

Date: dd/mm/yyyy	Revision Number	Summary of changes/ Reason for change	Section ref:	Author:

12.0 APPENDICES:

1. Appendix I:

Appendix I. Procedure Development Group/Stakeholders

Development Group Members/ Stakeholders	Approval Date
1 Louise Perris, CNEF, CHI Temple Street	
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