NSPKU The National Society for Phenylketonuria (UK) Ltd

Supporting patients with Phenylketonuria (PKU) in the surgery: a guide for GP administrative staff

What is PKU?

PKU is a rare genetic metabolic disorder affecting around 1:10,000 people in the UK, which is diagnosed by the newborn blood spot (heel prick) test shortly after birth. PKU affects the ability to metabolise the amino acid phenylalanine, which is found in protein. If PKU is not correctly treated in childhood, irreversible brain damage may occur causing severe intellectual disability. However, with the right treatment, people with PKU live healthy lives.

What is the treatment for PKU?

The treatment for PKU is a lifelong, very strict low protein diet with amino acid supplements. Patients cannot eat most foods such as meat, fish, eggs, soya, nuts, pulses or dairy foods such as milk and cheese. A lot of starchy foods such as bread, pasta and flour are also too high in protein. Many patients with PKU only tolerate 4g or 5g of protein per day which is equivalent to the protein found in one slice of bread.

Practical steps

To ensure that patients with PKU receive consistent product supply of the correct low protein foods and protein substitutes:

- Patients may be registered with more than one third party provider. The following companies have home delivery agreements and their staff have received training in the management of PKU:
  - **Vitaflo at Home** (Vitaflo and Fate products)
  - **Homeward** (Nutricia Metabolics products)
  - **Dial a Chemist** (Mevalia, Promin, Taranis and Metax products)
  - **HealthNet** (Cambrooke products)
- Identify on the home screen the name of the third party provider and the products they issue.
- Ensure that prompts/checks on the surgery IT systems, relating to the patients ACBS items, assist the processing of repeat prescriptions and home deliveries.

Please remember...

Patients with PKU have no choice and must rely on a number of ACBS items to manage their very rare condition. Unlike coeliac disease, these items cannot be bought elsewhere.

They will have a specialist metabolic dietitian who has expertise in PKU and can assist with any queries.

The help, understanding and support of GPs and administrative staff make a big difference to patients with PKU.

> "Patients frequently tell me that they find it difficult to obtain prescriptions for supplies of ACBS foods and protein substitutes. Their prescriptions may be cut, reduced, interrupted or continually questioned. This can cause stress and upset to families and individuals living with PKU. Lack of ACBS low protein foods and protein substitutes cause poor metabolic control and this damages the brain.
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> People with PKU have an unusually high number of repeat ACBS prescriptions due to the nature of their condition. I hope this guide will help medical administrative staff understand PKU."
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> *Prof. Anita MacDonald, Birmingham Children's Hospital*

> "Patients rely on a consistent supply of prescribed ACBS low protein foods for a large proportion of their diet including bread, pasta, flour, starch based sausage and burger mixes and snacks which are specially manufactured to be low in phenylalanine. They must also have ACBS prescribed protein substitutes which must be taken several times per day."
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> *Suzanne Ford RD, NSPKU Dietitian*

To help you, the NSPKU have produced guidance about the number of items of low protein products to prescribe each month. This is available on the NSPKU website.

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