

02/02/2017

Dear Simon,

Thank you for your letter dated 27th January 2017 in relation to my son ..... and the provision of low protein foods to patients with PKU. Your letter was in response to a letters I wrote to you in February 2016 & November 2016.

This response from the HSE does not appear to understand the rarity of inborn errors of metabolism and how this is a challenge for patients and their families. Equally this response does not address the challenges of manufacturing and distributing low protein foods. The HSE response consistently compares low protein products to gluten free. Quite simply there is no comparison.

PKU is an orphan disease. It is so rare that the likelihood of low protein products becoming available for consumption by the general population in shops, supermarkets or health shop is minimal. In fact, it would be very dangerous and irresponsible for a normal person to consume low protein products. However, gluten free products will not adversely affect a normal person's health in any way. I fail to see the comparison this HSE response is making to the availability of gluten free products in Ireland.

The HSE response suggests that suppliers must make their products available to Irish patients. PKU is an orphan disease. I fail to see how they can supply products like low protein bread and pasta through normal distribution channels when there are so few patients/ consumers who are spread out across all parts of Ireland. There simply isn't a demand for low protein products in the mainstream.

The HSE response states that new products have become available but suppliers have not sought reimbursement. This is quite simply untrue Simon. No new supplier has been given the opportunity to submit new products to the LTI list. There has not been a review in 5 years +. The HSE were not corresponding with these suppliers up to December 2016. The review process is just beginning now, January 2017. This information and the lack of HSE correspondence has been directly provided to the PKUAI from the low protein suppliers.

In your letter of 27th January 2017, you outline some of the criteria for reimbursement of medical grade foods specific to conditions such as PKU and HCU. Can you please provide further detail on what you mean by '..... The HSE has, in the past, made exceptional arrangements for staples such as bread. ....where a product is dietician- recommended and is less than an existing reimbursable item, it may be approved on a patient-specific basis'? What is the mechanism for doing so? I followed this procedure and applied to the HSE through the appropriate channel in early February 2016 having contacted Kate Mulvenna MPSI, Head of Pharmacy Function, Primary Care

Reimbursement Section. My application was completed by dieticians in Temple Street and sent to the HSE. I am still awaiting a response exactly 12 months later.

Simon, having perceived the previous HSE response as vague and inaccurate, I would like the following points addressed by the HSE:

What is the LTI budget spend on PKU products year on year (please provide data from the last 5 years)?

Who is responsible for maintaining the list?

How often do they meet?

What criteria are used in the assessment of new products for inclusion?

How is the decision made to remove products from the LTI list?

Where can we access reports relating to the assessment of products (other than FOI)?

How long does it take for an application to be assessed?

How does the number of products available here compare to other EU countries?

Where can we review which products have been assessed, are waiting to be assessed, and when those in waiting are going to be assessed?

How is the patient/ carer voice provided in the approval process? How can we, the end users give our input as to the value, or otherwise, of foods?

Is it acknowledged that an increased selection of foods does not necessarily equate to increased costs?

Before I conclude, I would like to make one further point Simon which I feel is important to stress. Living with PKU, a chronic illness is a constant daily challenge for us as a family and has had a significant impact on our quality of life. Managing Conor's condition offers serious daily challenges relating to his very strict diet restrictions. Conor has a 'classic' or severe strain of classic PKU and is therefore limited to only 4 regular grams of protein or exchanges per day (e.g. 1 exchange = 1 level tablespoon of oats/ 2 level tablespoons of peas) and restricts the intake of normal everyday protein foods such as: potatoes, bread, pasta, rice, cereals etc. The amount of protein that PKU patients can safely ingest does not increase as they grow. Therefore, this will be very challenging as he grows as we will have to rely completely on the very restricted selection of low-protein foods currently available within Ireland. As a result of PKU, if Conor does not strictly adhere to his diet, he is also at risk of many other health complications. Going off diet can cause many serious cognitive, neurological and psychological problems for PKU patients, which in

time, would put increased strain on our health system in the future. However, early intervention is the key and optimally treated PKU patients can live long healthy lives. Conor and other PKU sufferers have the potential to make an enormous contribution to society throughout his lifespan. I hope that with your help we can optimize outcomes for Conor, including his psychiatric outcomes, that may be jeopardized without access to appropriate care.

I would greatly appreciate the opportunity to meet with you and discuss the above further, to ensure a better future for Conor, and other PKU sufferers. If you require any further detail or information about PKU, please get in touch. I look forward to your reply.

Yours Sincerely,