

## **Adult Metabolic Diseases Clinic**

**Vancouver General Hospital**

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Mr. William V. Baker  
Commissioner - Chief Executive Officer of the CRA  
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555 MacKenzie Avenue  
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October 16, 2007

Dear Mr. Baker,

I am writing on behalf of the Canadian physicians who care for patients with genetic disorders affecting their metabolism. I am writing to request that the CRA allow the incremental costs of low protein products to be claimed as a medical expense, similar to the current situation with celiac disease and gluten free products.

There are many disorders affecting protein metabolism. The most common of these disorders is phenylketonuria (PKU) which affects 1/15 000 babies born in Canada each year. Other disorders affecting metabolism include defects of the urea cycle (UCD), other amino acid disorders such as maple syrup urine disease and organic acidemias (OA). In all of these disorders affecting protein metabolism, the patient is unable to break down part or all of the protein components, leading to accumulation of toxic metabolites. This accumulation can lead to mental retardation (as in the case of PKU), birth defects in women who have the disorder and become pregnant (PKU), or acute decompensation leading to coma and death (UCD and OA). Patients with these disorders require restriction of dietary protein to prevent accumulation of toxic metabolites. However, restriction of dietary protein is not enough to prevent decompensation as, if the patient does not consume a diet which is adequate in other nutritional components, the patient will break down their own muscle protein stores. Thus, the dietary therapy involves both restriction of the offending component of protein and supplementation with other foods to meet nutritional demands.

As part of these demanding diets, patients consume medical formulae to meet some of their nutritional needs. These formulae are considered essential to their care and are covered under provincial health plans in all provinces. However, patients also require the use of low protein foods to help them meet their daily caloric requirements. These low protein foods are not covered under many provincial health plans, leading to regional disparities in access to these products. Further, these low protein foods are extremely expensive. An analysis of the incremental cost of the low protein diet in a typical patient with PKU, based on a national survey of food costs, has been attached to this letter. In that analysis (see summary table 2.17), the incremental monthly cost of the low protein diet when compared to diet based on Canada Food Guide recommendations ranged from \$80.64-\$291.81 depending on the age and gender of the patient and his/her tolerance for phenylalanine.

I would like to suggest that the CRA consider including the incremental costs of purchasing low protein products as a medical expense on the individual T1 tax return for individuals with disorders where the use of low protein products is medically indicated. This would be similar to the

exemption that is currently available to the patients with celiac disease who can claim incremental costs for gluten free products. I would propose that similar documentation be required for individuals prescribed low protein products (including a letter from a medical practitioner confirming that the individual has a disorder for which low protein products are a required part of therapy and a summary of each item purchased).

I have enclosed a table which describes regional disparity in the funding of low protein products based on 2007 Canadian data. I have also enclosed the recent analysis of the incremental costs associated with the low protein diet in patients with PKU. Similar analyses of incremental costs associated with low protein diets are not available for patients with other disorders of protein metabolism due to the rarity of these disorders. However, the costs would not be expected to exceed those quoted for PKU.

I recognize that, in order to evaluate this fully, you will need a list of the disorders for which low protein products are prescribed as part of medically indicated therapy. A complete list of the various genetic disorders affecting protein metabolism and their prevalence would need to be compiled. Data on prevalence of these disorders will be available on review of newborn screening programs. However, for your initial consideration, I am also including a British Columbia publication where the actual BC prevalence of genetic disorders affecting metabolism is outlined. If you look at Table 1 in that document, you see that the first 4 disorders listed (amino acid diseases excluding PKU, PKU, UCD, and OA) have a combined incidence of 20.7/100 000 live births and this figure will include all of the most common disorders. Thus, the economic impact of this proposal will be considerably smaller than that for celiac disease which has already been approved by the CRA.

I have enclosed faxed copies of signatures from other physicians across the country who support the proposal outlined in this letter. I would be happy to work further with the CRA to provide information necessary for further evaluation of this proposal. I would strongly urge the CRA to consider the incremental costs of low protein products as a medical expense for patients who require low protein diets due to genetic disorders affecting their metabolism.

Yours sincerely,

Dr. Sandra Sirrs MDFRCPC  
Medical Director, Adult Metabolic Diseases Clinic