Data in Brief 13 (2017) 377–384

Contents lists available at ScienceDirect

Data in Brief

journal homepage: www.elsevier.com/locate/dib

Data Article

Dietary amino acid intakes associated with a low-phenylalanine diet combined with amino acid medical foods and glycomacropeptide medical foods and neuropsychological outcomes in subjects with phenylketonuria

Bridget M. Stroup [a], Sangita G. Murali [a], Nivedita Nair [a], Emily A. Sawin [a], Fran Rohr [b], Harvey L. Levy [b], Denise M. Ney [a,*]

[a] Department of Nutritional Sciences, University of Wisconsin-Madison, WI, United States
[b] Division of Genetics and Genomics, Boston Children's Hospital, Harvard Medical School, Boston, MA, United States

ARTICLE INFO

Article history:
Received 6 April 2017
Received in revised form
10 May 2017
Accepted 1 June 2017
Available online 7 June 2017

Keywords:
Tyrosine
Leucine
Arginine
Executive function
Delis-Kaplan Executive Function System
Cambridge Neuropsychological Test Automated Battery

ABSTRACT

This article provides original data on median dietary intake of 18 amino acids from amino acid medical foods, glycomacropeptide medical foods, and natural foods based on 3-day food records obtained from subjects with phenylketonuria who consumed low-phenylalanine diets in combination with amino acid medical foods and glycomacropeptide medical foods for 3 weeks each in a crossover design. The sample size of 30 subjects included 20 subjects with classical phenylketonuria and 10 with a milder or variant form of phenylketonuria. Results are presented for the Delis-Kaplan Executive Function System and the Cambridge Neuropsychological Test Automated Battery; the tests were administered at the end of each 3-week dietary treatment with amino acid medical foods and glycomacropeptide medical foods. The data are supplemental to our clinical trial, entitled "Glycomacropetide for nutritional management of phenylketonuria: a randomized, controlled, crossover trial, 2016 (1) and "Metabolomic changes demonstrate reduced bioavailability of tyrosine and altered metabolism of tryptophan via the kynurenine pathway with

DOI of original article: http://dx.doi.org/10.1016/j.ymgme.2017.04.003
* Correspondence to: University of Wisconsin-Madison, Department of Nutritional Sciences, 1415 Linden Drive, Madison, WI 53706 Fax: +1 608262 5860.
E-mail address: ney@nutrisci.wisc.edu (D.M. Ney).

http://dx.doi.org/10.1016/j.dib.2017.06.004

ingestion of medical foods in phenylketonuria, 2017 (2). This data has been made public and has utility to clinicians and researchers due to the following: 1) This provides the first comprehensive report of typical intakes of 18 amino acids from natural foods, as well as amino acid and glycomacropeptide medical foods in adolescents and adults with phenylketonuria; and 2) This is the first evidence of similar standardized neuropsychological testing data in adolescents and adults with early-treated phenylketonuria who consumed amino acid and glycomacropeptide medical foods.

Specifications Table

Subject area	*Biology, Medicine*
More specific subject area	*Inherited Metabolic Disorders*
Type of data	*Figure (study design), Tables (dietary amino acid intakes, neuropsychological testing outcomes)*
How data was acquired	*Assessment of dietary intake of amino acids and neuropsychological function in patients with PKU*
Data format	*Analyzed data, mean ± SD, median (25th–75th percentile)*
Experimental factors	*Data of subjects with PKU enrolled in Clinical trial at Waisman center, Madison, WI and Boston Children's hospital, Boston, MA.*
Experimental features	*Randomized Crossover Clinical Trial*
Data source location	*Madison, Wisconsin, USA*
Data accessibility	*The data are accessible within the article*

Value of the data

- The data presented are the first comparison of how ingestion of medical foods comprised primarily of single amino acids or intact protein from glycomacropeptide (a 64-amino acid glycophosphopeptide isolated from cheese whey) affect the dietary intake profile of amino acids.
- The dietary intake of 18 amino acids provides useful information to clinicians and researchers related to typical amino acid intake of individuals with phenylketonuria.
- The data from the standardized neuropsychological tests can be compared with pharmacological studies using these same tests to contrast the effectiveness of dietary management with pharmacological treatment in subjects with phenylketonuria [3,4].
- These data are useful to clinicians and researchers evaluating the safety and efficacy of glycomacropeptide medical foods in the nutritional management of phenylketonuria.

1. Data

Summary data for dietary intake of amino acids and assessment of neuropsychological and executive function are presented for subjects with phenylketonuria enrolled in a randomized, controlled, crossover trial conducted from November 2010 to July 2015 [1,2]. These data are herein reported for the first time (Tables 1-3). The trial is registered at www.clinicaltrials.gov as NCT01428258 .