

Methods: In the present study, we examined whether 6.7g of leucine enriched essential amino acid mixture (LEU/EAA) induced more protein anabolism than the same dose of a balanced mixture of essential and non-essential amino acids (TAA) as present in Whey protein in 14 clinically stable pediatric patients with CF (mean age: 15±2y, FEV1: 85±22%pred.). Fifteen grams of maltodextrin was added as a source of carbohydrates. Whole body protein synthesis (PS) and breakdown (PB) and net PS (= PS-PB) were measured by the combined infusion of the stable isotopes L-[ring-2H5]phenylalanine and L-[ring-2H2]tyrosine. Myofibrillar PB, as indirect marker of skeletal muscle protein breakdown was measured by L-[2H3]-3-Methylhistidine and collagen PB by [2H]-hydroxyproline. The isotope enrichment values and amino acid concentrations were measured in arterialized-venous plasma, and Fat-free mass (FFM) was assessed using Dual-energy X-ray absorptiometry. Presence of nutritional failure in the CF group was defined according to the criteria BMI percentile < 10th and/or FFM-index < 5th percentile. Statistics was done using (un)paired t-tests when appropriate.

Results: Whole-body PS ($p<0.05$) but not PB, myofibrillar PB or collagen PB was significantly stimulated after intake of the LEU/EAA mixture as compared to the TAA mixture, resulting in 70% higher values for net PS ($P<0.001$). Stratification of the CF group in nutritional failure ($n=7$) vs. normal nutritional status ($n=7$), showed no difference in (net) PS and PB values (whole body, myofibrillar or collagen) after LEU/EAA intake indicating comparable elevated values for net PS as compared to the TAA mixture in both groups with CF.

Conclusion: A dietary Leu/EAA mixture has a significantly higher anabolic potential than a balanced mixture of amino acids in pediatric patients with CF, independent of the presence of nutritional failure. This indicates that even patients with CF characterized by nutritional failure are responsive to amino acid modulation.

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DIETARY ESSENTIAL AMINO ACIDS ARE HIGHLY ANABOLIC IN PEDIATRIC PATIENTS WITH CYSTIC FIBROSIS CHARACTERIZED BY NUTRITIONAL FAILURE

Engelen, M.P.¹; Com, G.²; Wolfe, R.R.¹; Deutz, N.E.^{1,2} 1. *Geriatrics, Center for Translational Research in Aging & Longevity, University of Arkansas for Medical Sciences, Little Rock, AR, USA*; 2. *Pediatric Pulmonology, Arkansas Children's Hospital, Little Rock, AR, USA*

Background: Despite the pivotal role of nutrition in CF, current approaches to treat muscle loss by nutritional intervention have only been partially successful. To circumvent diminished protein digestibility due to pancreatic insufficiency in CF, (semi) elemental diets containing free amino acids are advised. We and others have previously found in diseases and conditions associated with muscle wasting that mixtures of essential amino acids (EAA) are more effective in stimulating whole body and muscle protein synthesis than equal amounts of balanced mixtures of EAA and non-essential amino acids as present in dietary (milk) protein. However data on the anabolic properties of EAA in CF are lacking.