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NUTRITIONAL FAILURE IS ASSOCIATED WITH ENHANCED WHOLE BODY PROTEIN TURNOVER AND CATABOLISM IN PEDIATRIC PATIENTS WITH CYSTIC FIBROSIS

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Background: Malnutrition commonly occurs in pediatric patients with cystic fibrosis (CF) negatively influencing their quality of life and survival. Recently we have shown that a substantial number of pediatric patients with CF are characterized by nutritional failure based on low body mass index (BMI) and/or fat-free mass index (FFMI). It is unclear whether these CF patients have higher protein turnover and breakdown that relate to more susceptibility for protein loss and to more energy costs.

Methods: In the present study, we examined in the postabsorptive state whether whole body protein breakdown (PB) and synthesis (PS) differ between 14 stable pediatric patients with CF and 17 healthy young controls. Whole-body PB, PS, and net PB (= PB-PS) 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. In arterialized-venous plasma, the isotope enrichment values and amino acid concentrations were measured. FFM was measured 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 FFMI < 5th percentile. Lung function (as reflected by forced expiratory volume in 1 second (FEV1)) of each CF patient was recorded. Statistics was done using ANOVA and t-tests when appropriate.

Results: Whole-body PS, PB ($p< 0.001$) and net PB ($p<0.05$), but not myofibrillar PB was higher in the CF group (mean age: 15±2y, FEV1: 85±22%pred.) than in the control group. Stratification of the CF group into nutritional failure (n=7) vs. normal nutritional status (n=7) resulted in higher values for whole body PS and PB ($p<0.05$) in the group with nutritional failure. Net PB, myofibrillar PB, FEV1 and age were not different between the CF groups.

Conclusion: The present study reveals that nutritional failure in CF is associated with elevated levels for whole body protein turnover and increased net catabolism in pediatric patients with CF, indicating that specific nutritional modulation is warranted to target the abnormalities in protein and amino acid metabolism in CF.