New ways of defining protein and energy relationships in inborn errors of metabolism.

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Abstract
Dietary restrictions required to manage individuals with inborn errors of metabolism (IEM) are essential for metabolic control, however may result in an increased risk to both short and long-term nutritional status. Dietary factors most likely to influence nutritional status include energy intake, protein quality and quantity, micronutrient intake and the frequency and extent to which the diet must be altered during periods of increased physical or metabolic stress. Patients on the most restrictive diets, including those with intakes consisting of low levels of natural protein or those with recurrent illness or frequent metabolic decompensation carry the most nutritional risk. Due to the difficulties in determining condition specific requirements, dietary intake recommendations and nutritional monitoring tools used in patients with IEM are the same as, or extrapolated from, those used in healthy populations. As a consequence, evidence is lacking for the safest dietary prescriptions required to manage these patients long term, as tolerance to dietary therapy is generally described in terms of metabolic stability rather than long term nutritional and health outcomes. As the most frequent therapeutic dietary manipulation in IEM is alteration in dietary protein, and as protein status is critically dependent on adequate energy provision, the use of a Protein to Energy ratio (P:E ratio) as an additional tool will better define the relationship between these critical components. This could accurately define dietary quality and ensure that not only an adequate, but also a safe and balanced intake is provided.

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KEYWORDS: Body composition; Inborn errors of metabolism (IEM); Nutritional outcomes; Protein restricted diet; Protein:energy ratio (P:E ratio)

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