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# Dietary practices in methylmalonic acidaemia: a European survey

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## Abstract

### Background

The dietary management of methylmalonic acidemia (MMA) is a low-protein diet providing sufficient energy to avoid catabolism and to limit production of methylmalonic acid. The goal is to achieve normal growth, good nutritional status and the maintenance of metabolic stability.

### Aim

To describe the dietary management of patients with MMA across Europe.

### Methods

A cross-sectional questionnaire was sent to European colleagues managing inherited metabolic disorders (IMDs) (n=53) with 27 questions about the nutritional management of organic acidaemias. Data were analysed by different age ranges (0–6 months; 7–12 months; 1–10 years; 11–16 years; >16 years).

## Results

Questionnaires were returned from 53 centres. Twenty-five centres cared for 80 patients with MMA vitamin B12 responsive (MMAB12r) and 43 centres managed 215 patients with MMA vitamin B12 non-responsive (MMAB12nr). For MMAB12r patients, 44% of centres (n=11/25) prescribed natural protein below the World Health Organization/Food and Agriculture Organization/United Nations University (WHO/FAO/UNU) 2007 safe levels of protein intake in at least one age range. Precursor-free amino acids (PFAA) were prescribed by 40% of centres (10/25) caring for 36% (29/80) of all the patients. For MMAB12nr patients, 72% of centres (n=31/43) prescribed natural protein below the safe levels of protein intake (WHO/FAO/UNU 2007) in at least one age range. PFAA were prescribed by 77% of centres (n=33/43) managing 81% (n=174/215) of patients. In MMAB12nr patients, 90 (42%) required tube feeding: 25 via a nasogastric tube and 65 via a gastrostomy.

## Conclusions

A high percentage of centres used PFAA in MMA patients together with a protein prescription that provided less than the safe levels of natural protein intake. However, there was inconsistent practices across Europe. Long-term efficacy studies are needed to study patient outcome when using PFAA with different severities of natural protein restrictions in patients with MMA to guide future practice.

This article offers supplementary material which is provided at the end of the article.

**Keywords:** methylmalonic acidaemia; natural protein; precursor-free amino acids; protein-restricted diet

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