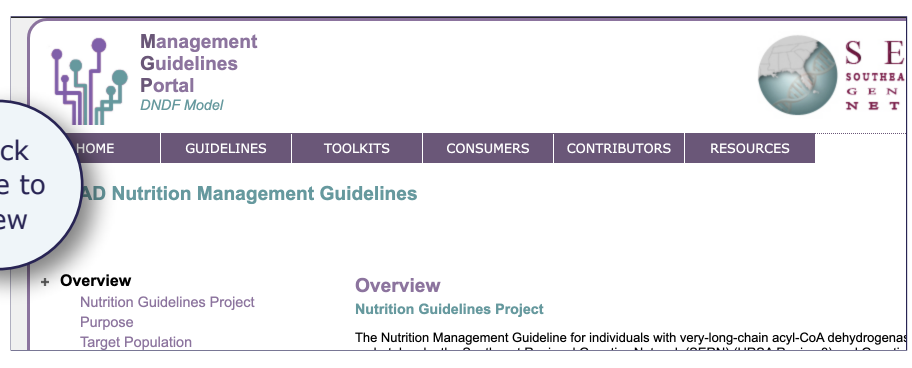
One of the groups belonging to the lipid disorders is the FAODs. The dietary management of FAOD presents many challenges. Here we present key papers that discuss and offer guidance on some of the challenges we face.

Management Guidelines And Recommendations

VLCAD Nutrition Management Guidelines

- **SERN/GMDI 2019**

In 2019, SERN and GMDI published an updated Nutrition Management Guideline for individuals with VLCAD deficiency.

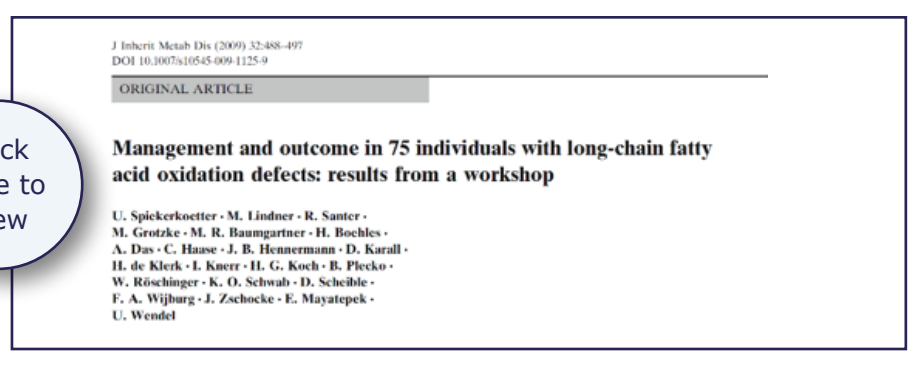


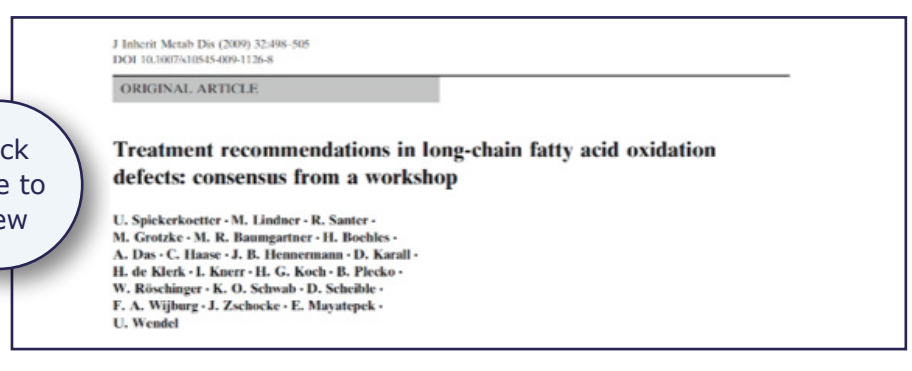
The guideline was first published back in 2008. The updated version gives a comprehensive overview and main updates to the recommendations include:

- Updated recommendations for total fat, LCT and MCT intake with more detailed categorisation according to age and disorder severity
- Updated recommendations for EFA and DHA supplementation
- Recommendations for breast-feeding in symptomatic and asymptomatic infants
- Updated recommendation for the use of corn starch

Spiekerkoetter et al 2009

Two papers from Spiekerkoetter's group in 2009 give consensus recommendations based on the observation of 75 patients, together with expert opinion. Although older than the GMDI guidelines, these recommendations consider a wider patient group, offering disease-specific recommendations for VLCAD, LCHAD, TFP complex, CPTI and CPTII.

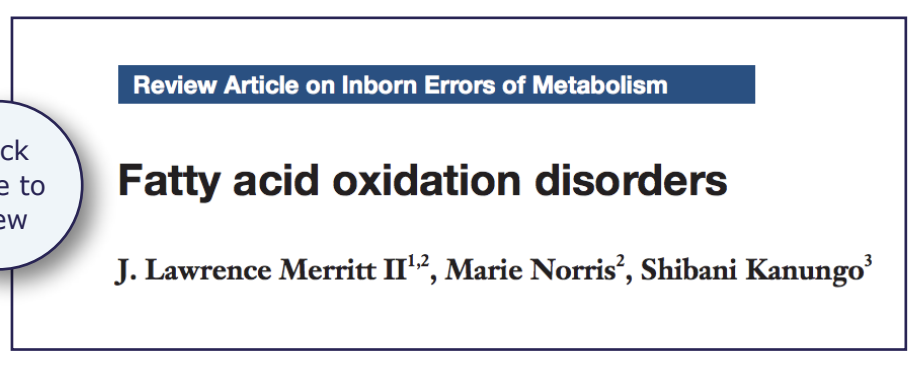




The main recommendations relating to dietary management include:

- Suggested maximum fasting periods categorised by age
- LCT restriction and MCT supplementation for VLCAD, LCHAD and TFP complex
- EFA and DHA supplementation

Merritt et al 2018

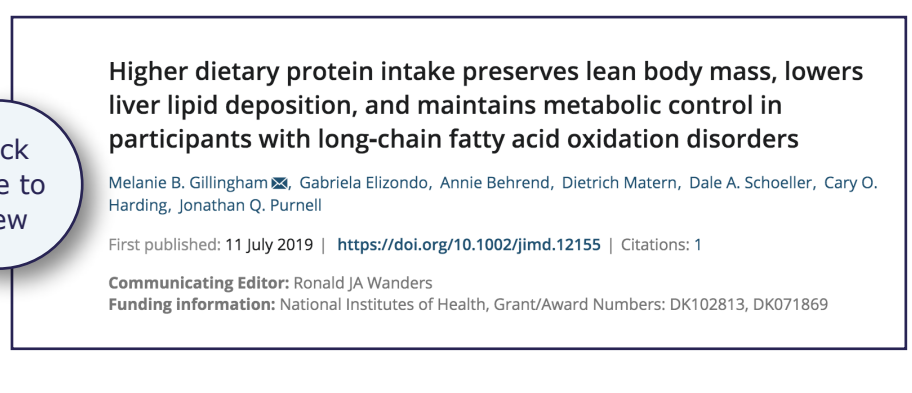


The article discusses nutritional management of each disorder, recognising different practices and includes an extensive reference list.
Wider considerations for dietary management are addressed, including protein provision and management of exercise.

The paper concludes with an appeal for more randomised controlled trials to evaluate and progress therapies.

Protein Intake In Fatty Acid Oxidation Disorders

With fat intakes limited, higher carbohydrate intakes are often used to ensure an adequate energy intake. Building on previous work, in this 2019 paper Gillingham and colleagues examined the impact of a higher protein diet compared to one higher in carbohydrate.



Increased lean body mass and a trend towards reduced liver fat deposition were found in children and young adults taking a higher protein to carbohydrate ratio. These findings contribute to wider goals of a balanced diet and a healthy and active lifestyle.

High MCT Formula In Fatty Acid Oxidation Disorders

The clinical diversity of LC-FAOD makes it difficult to find a formula to suit all. MacDonald and colleagues from Birmingham Children's Hospital are first to report on the use of an MCT-based formula designed specifically for LC-FAOD.

The study population includes children with VLCAD, LCHAD and CACT deficiencies and reports on tolerance, biochemical measurements, muscle pain and requirement for supplementation over a 21-day period. The authors conclude that the formula appears safe and well-tolerated, requiring no further supplementation of LCPUFAs in the study group.

Abbreviations

GMDI	Genetic Metabolic Dietitians International
FAOD	Fatty Acid Oxidation Disorder(S)
LC-FAOD	Long Chain Fatty Acid Oxidation Disorder(S)
CACT	Carnitine Acylcarnitine Translocase
CPT I	Carnitine Palmitoyl Transferase I
CPT II	Carnitine Palmitoyl Transferase Ii
DHA	Docosahexaenoic Acid
EFA	Essential Fatty Acid
LCHAD	Long Chain 3-Hydroxyacyl-Co-A Dehydrogenase
LCPUFA	Long Chain Polyunsaturated Fatty Acid
LCT	Long Chain Triglyceride
MCT	Medium Chain Triglyceride
SERN	Southeast Regional Genetics Network
TFP complex	Trifunctional Protein Complex
VLCAD	Very Long Chain Acyl-CoA Dehydrogenase