Introduction and Purpose

Long-chain fatty acid oxidation disorders (LC-FAOD) are rare inborn errors of metabolism for which dietary management is the cornerstone of treatment. This study was conducted to understand knowledge, attitudes, barriers, and experience related to managing patients with LC-FAOD in order to determine areas of educational need for future continuing education to address.

Methodology

1. A survey instrument, including patient case scenarios with two of the most common forms of LC-FAOD (CPT-II and VLCAD), was designed to understand knowledge, attitudes, barriers, and experience related to managing patients with LC-FAOD.
2. The survey was developed and tested in collaboration with a dietitian experienced with managing LC-FAOD patients.
3. The survey was distributed via email to 672 US-practicing dietitians during September 2019. Fifty-two dietitians completed the survey, of which 20% had previously managed a patient with LC-FAOD.
4. Descriptive data analysis and open-ended data coding was performed.

Respondent Demographics

<table>
<thead>
<tr>
<th>Practice Location</th>
<th>Suburban</th>
<th>Rural</th>
<th>Urban</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients seen per week (mean)</td>
<td>29 patients</td>
<td>19 years</td>
<td></td>
</tr>
<tr>
<td>Years since earning dietitian registration/nutrition license (mean)</td>
<td>19 years</td>
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</tbody>
</table>

Dietary recommendations

Case: An 18-month-old with very long chain acyl CoA dehydrogenase deficiency

Dietary recommendations to prevent future acute episodes and complications of disease:
- Frequent meals and/or avoidance of extended fasting: 50%
- Medium chain triglyceride supplementation: 48%
- High-carbohydrate, low fat diet: 42%
- Carbohydrate supplementation: 31%
- Essential fatty acid supplementation: 31%
- Increased fluid intake: 27%
- Cornstarch or multigrain in the evening or between meals: 19%
- Vitamin/mineral supplementation: 17%
- Increased caloric intake: 15%
- Semi-elemental or elemental supplement: 4%
- No changes to diet are needed: 2%
- Deferral dietary recommendations to another specialist: 25%
- Other: 6%

75% reported they would make dietary recommendations rather than referring to another specialist. Of those making recommendations, the majority would advise avoidance of fasting: a low-fat diet, medium-chain triglyceride oil, or carnitine.

Familiarity with LC-FAOD Pathophysiology

Where does fatty acid oxidation primarily take place?
- Cytosol: 19%
- Endoplasmic reticulum: 4%
- Golgi apparatus: 4%
- Mitochondrial matrix: 50%
- Unsure: 23%

Which of the following is produced by the oxidation of odd-chain fatty acids that is NOT produced by the oxidation of even-chain fatty acids?
- Acetyl CoA: 25%
- Butyryl CoA: 2%
- Palmitoyl CoA: 4%
- *Propionyl CoA: 21% (unsure)

Many dietitians are unfamiliar with the normal processes of fatty acid metabolism, suggesting that future education should include concepts relating to both physiology and pathophysiology of LC-FAOD.

Patient Education

Case: 15-year-old with newly diagnosed carnitine palmitoyltransferase II deficiency

<table>
<thead>
<tr>
<th>Would discuss with following patient/parent:</th>
<th>Diet habits</th>
<th>Food preparation</th>
<th>Strategies for eating away from home</th>
<th>Strategies for preventing hypoglycemia</th>
<th>Signs/symptoms to watch for</th>
<th>Disease impact on the Krebs cycle</th>
<th>Disease pathophysiology</th>
<th>Disease genetics and/or heredity</th>
<th>Prognosis</th>
<th>Investigational therapies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Percentage</td>
<td>100%</td>
<td>100%</td>
<td>100%</td>
<td>98%</td>
<td>54%</td>
<td>52%</td>
<td>48%</td>
<td>44%</td>
<td>25%</td>
<td>14%</td>
</tr>
</tbody>
</table>

Less than half of dietitians would discuss disease pathophysiology, impact on metabolism, and genetic basis with the patient diagnosed with LC-FAOD, which may be a reflection of lack of knowledge surrounding these topics.

Barriers in Managing Patients with LC-FAOD

| Perceived significance of barriers to optimal management of patients with LC-FAOD |
|----------------------------------|-------------|----------------|----------------|----------------|----------------|----------------|----------------|----------------|
| Lack of effective therapies | 52% | Lack of management guidelines | 59% | Patient/parent difficulty in adhering to dietary recommendations | 31% | Lack of education on LC-FAOD during dietitian training | 31% | 

The most significant barrier to dietitians in optimally managing patients with LC-FAOD is the lack of education on LC-FAOD during dietitian training.

Continuing Education

Do you typically attend national meetings? Yes: 52%
Do you access online continuing education? Yes: 98%

Which of the following is your preference for the content of continuing education?
- Content directed solely to dietitians: 64%
- Content directed to a multidisciplinary team including physicians: 35%
- Other: 2%

LC-FAOD Management

How important are each in making dietary recommendations for patients with LC-FAOD?
- A standard protocol used in my practice: 12%
- The patient’s overall health and history of hospitalizations: 44%
- The patient’s/parent’s level of interest in disease management: 41%

Familiarity with emerging therapies for LC-FAOD

<table>
<thead>
<tr>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bexarotene</td>
<td>1</td>
<td>Gene therapy</td>
<td>1</td>
<td>Triptandrin</td>
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</table>

Dietitians report placing more importance on an individualized approach to dietary management than on following a standard protocol. There is low familiarity with emerging therapies for LC-FAOD amongst dietitians.

Implications and Conclusion

Dietitians play a critical role in helping patients manage LC-FAOD through a modified diet and supplementation. However, dietitians report receiving limited training on LC-FAOD and many do not have experience managing patients with LC-FAOD. This study identified areas for future education directed toward dietitians, including awareness of disease pathophysiology and nutritional management to prevent episodes of metabolic decompensation. Due to small sample size, data were not evaluated based on those who have managed LC-FAOD versus those who have not. However, results support that continuing education should be designed to address differences in dietitians’ learning preferences including online education.