Treatment and Care of Cystic Fibrosis Related Diabetes (CFRD): A Team Approach

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Diane Paralusz, RD, CSP
Margaret Potter, PharmD

Cystic Fibrosis: Background

- Genetic disorder
  - Exocrine glands
    - Lungs, pancreases, liver, reproductive organs
  - Medication: 50 – 60 pills daily
  - Respiratory: treatments 2 hours daily (more when sick)
- Nutrition
- Exercise
- Daily living
- Psychosocial

CFRD: Background

- 15-20% adolescents
- 40-50% adults
- Varied etiology
  - Insulin insufficiency
  - Insulin resistance
**CFRD: Background**
- **Onset:**
  - Insidious vs. Acute
- **Complications:**
  - Microvascular: YES
  - Macrovascular: NO
- **Multidisciplinary Team**

**Screening**
- Annual Screening
  - Start age 10
  - Preconception & Gestational
  - Pre-transplantation
- Oral Glucose Tolerance Test (OGTT)
  - Stable baseline health
  - Exacerbation
  - Continuous feeding

**Not Recommended**
- Urine glucose
- Random glucose
- Fasting glucose
- A1c
- Continuous Glucose Monitor

**Multidisciplinary Team**
- A1c
- Microvascular screening
- Lipid profile
- "C-peptide prn"

**Education: Self Management**
- CHO counting
- SMBG TID
- Nutrition
- Exercise

**Insulin Therapy**
- Impaired Glucose Tolerance (IGT)
  - Rapid acting/Short Acting Insulin
- Impaired Fasting Glucose (IFG)
  - Basal Insulin: Analog
- Coverage for Overnight Feeding
  - 4 hour feeding
    - Regular
  - 6 to 8 hour feeding
    - NPH

**ADA, 2016; Moran et al., 2010**
Complications

- Acute:
  - Hypoglycemia
  - Decreased Glucagon production
- Chronic:
  - Nephropathy
  - Retinopathy
  - Gastroparesis
  - Direct impact on pulmonary function and overall mortality
    - Weight loss/Malnutrition
    - Protein Catabolism
    - Infection

Anderson et al., 2006; Caraher et al., 2014; Moran et al., 2010

Nutrition and CF-Related Diabetes

- Goals:
  - to achieve/maintain optimal nutritional status and normalize blood glucose to preserve lung function
  - Sustained growth and weight gain in children and weight gain/maintenance in adult patients with CF

Nutrition and Cystic Fibrosis

- The Cystic Fibrosis Foundation (CFF) recommends:
  - Goal BMI: >50% tile for ages 2-20 years old
  - Goal BMI: >23 for males >20 years
  - Goal BMI: >22 for females >20 years

Nutritional Issues in Cystic Fibrosis

- Patients with cystic fibrosis are at high risk for malnutrition secondary to:
  - Increase energy expenditure (progressive lung disease)
  - Reduced absorption (pancreatic insufficiency, small intestinal bacteria overgrowth, CF-related liver disease)
  - Inadequate intake (GERD, constipation, gastroparesis, sinusitis, fatigue, behavioral)
  - Increased energy loss (CF-related diabetes, posttussive emesis)

Dietitian’s Role

Diane Paralusz, RD, CSP

Nutrition Management for Cystic Fibrosis-Related Diabetes (CFRD)

- Objectives:
  - Participants will identify two differences between the nutrition management of CF Related Diabetes and other types of diabetes
Nutrition and CF-Related Diabetes

- Direct correlation between nutritional status, BMI, lung function (FEV1), and survival
- CF-Related Diabetes associated with decline in nutritional status and lung function
- Psychological Impact
  - Emotional and psychological burden of a second chronic illness


Nutrition Management for CFRD

<table>
<thead>
<tr>
<th>Nutrient</th>
<th>CF-Related Diabetes</th>
<th>Type 1 &amp; 2 Diabetes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calories</td>
<td>1.2-1.5 DRI or more individualized based on weight gain, growth, lung function</td>
<td>As needed for growth, maintenance, weight loss</td>
</tr>
<tr>
<td>Carbohydrate</td>
<td>Individualized; 40-50% total calories</td>
<td>Individualized; 45-50% total calories</td>
</tr>
<tr>
<td>Fat</td>
<td>25-30% total calories</td>
<td>30% total calories or less</td>
</tr>
<tr>
<td>Protein</td>
<td>1.2-2.0 g/kg for age, no reduction for nephropathy</td>
<td>0.8-1 g/kg for age, no reduction for nephropathy</td>
</tr>
<tr>
<td>Sodium</td>
<td>&gt;4000 mg/day; liberal, high salt diet</td>
<td>&lt;2300 mg/day</td>
</tr>
<tr>
<td>Vitamins, Minerals</td>
<td>CF-specific vitamin/mineral supplement</td>
<td>Individually based on diet or deficiency noted</td>
</tr>
</tbody>
</table>

Nutrition Management of CF-Related Diabetes

- Dietitian’s role
  - Comprehensive nutrition assessment
  - Evaluate anthropometric data (weight, height, BMI)
  - Obtain diet history
  - Biochemical
  - Clinical (malabsorption, vitamin/mineral deficiencies, CF-related diabetes, small intestinal bowel bacteria overgrowth)
  - Intervention
  - Education/Counseling

Nutrition Management of CFRD

- Diet:
  - High calorie/protein/fat, well balanced diet. Encouraged to spread carbohydrates throughout the day.
  - Carbohydrate counting ideal to optimize glycemic control.
  - Limit/avoid concentrated sources of sugar with little nutritional value.
  - Oral supplements, enteral feedings: no restriction; utilize 1.5-2 calorie/mL nutrient dense formulas.

Discharge Pharmacist’s Role
Margaret Potter, PharmD

Pancreatic Enzymes

- Inhibited exocrine function of pancreas
  - Inability to absorb fat in meals well → accelerate carbohydrate absorption
- Need to optimize pancreatic enzyme replacement therapy (PERT)
  - With meals, snacks, tube feeds
  - Lipase/protease/amylase
  - Brand names: Creon®, Zenpep®, Ultresa®, Pancreaze®
Role of Oral Diabetes Agents

- Not FDA approved
- Insulin drug of choice

Brennan et al., 2015

New Medications

- Ivacaftor (Kalydeco®)
  - With high fat meals
  - Potentiator
  - DDI with CYP3A4
- Ivacaftor/Lumacaftor (Orkambi®)
  - Potentiator/ corrector

Discharge Pharmacist Program

- New Medications
- Safe Discharge Home

New Medications

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Discharge Pharmacist Workflow

1. Monday-Sunday 0800 to 1830
   - RN receives order to d/c patient home
2. Patients with any of the following: COPD, CHF, PNA, AMI, ESRD, & DM > 5 scheduled medications
3. Pharmacist will educate patient on medications
   - Coordinate prescription preparation with outpatient pharmacy

Safe Discharge Home

- Insurance coverage
- Diabetes supply protocol
  - Test strips
  - Lancets
  - Glucometer
  - Syringes or pen needles
Follow Up Care

- Call patient 3-5 days post discharge
- Confirm medication pick up
- Tolerating medications
- Confirm follow up appointments

Pharmacist Interventions

- 2851 patients discharged
- 1961 patients discharged home
- 1249 patients seen by pharmacists
- 285 patients with interventions
- Interventions made in 23% of the patients seen

Results: High-Risk Characteristics

- Total of 390 interventions made on 285 patients

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<th>High-Risk Characteristics</th>
<th>No. (%)</th>
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<tr>
<td>5 or more scheduled medications</td>
<td>219 (56%)</td>
</tr>
<tr>
<td>Diabetes</td>
<td>179 (46%)</td>
</tr>
<tr>
<td>End-Stage Renal Disease</td>
<td>46 (12%)</td>
</tr>
<tr>
<td>Chronic Obstructive Pulmonary Disease</td>
<td>25 (6%)</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>7 (2%)</td>
</tr>
<tr>
<td>Congestive Heart Failure</td>
<td>6 (2%)</td>
</tr>
<tr>
<td>Multiple High-Risk Characteristics</td>
<td>148 (38%)</td>
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Results

November 2013

- 30-Day ED Visit LBM
  - With Pharmacist: 150, Without Pharmacist: 109
  - P=0.002

- 30-Day Readmission LBM
  - With Pharmacist: 142, Without Pharmacist: 100
  - P=0.002
**HCAHPS Scores – Care Transitions**

- **2012 – Without Pharmacist**
  - Average = 39.2

- **2013 – With Pharmacist**
  - Average = 55.9

**Plan…**

- **Diabetes Management**
  - Nutritional assessment
  - Appointments
  - Skill/Counting Compliance

- **Care Transitions**
  - Discharge
  - Diabetes supplies
  - Insulin supplies

**Case Study**

- **Linda Kerr, DNP, RN, FNP-BC, CDE**
- **Diane Paralusz, RD, CSP**
- **Margaret Potter, PharmD**

**Darren**

- **Age 25**

**Medical History**

- CF diagnosis (infant)
- CFRD diagnosis (age 18)
- Malnutrition
- Gastroparesis
- Chronic sinusitis
- Pulmonary function

**Insulin**

- Basal, bolus, correction, overnight coverage for GT feeds

**Overnight GT feeds**

- **The 4th meal**
  - 2 Kcal/mL formula @ 250 mL/hr x 4 hours
  - 216 carbs (54 Cho grams/hour)

**Diet**

**Questions**

- **Hospitalizations**
  - 2014: 4 admissions
  - 2015: 6 admissions

- **Clinic visits**
  - 2014: every 3 months
  - 2015: every 2 months
  - 2016: every 1–2 months

**References**