What Every Dietitian Needs to Know about Genetic Sucrase-Isomaltase Deficiency (GSID)

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Clinical Science Liaison  QOL Medical, LLC

Introduction (Don’t Worry—This Is Not A Test)

- How many have heard of Genetic Sucrase-Isomaltase Deficiency (GSID)?
- How many are familiar with foods high in sucrose?
- What about foods that are high in starch?
- GSID is not something we learned about in school.
- GSID is not something most physicians are familiar with.
- But, there are patients suffering with GSID.
- And, I believe dietitians are in the perfect place to help identify these patients, recommend further diagnostic testing, and provide nutrition education to those with GSID.

Disclosures

- Employed by QOL Medical, LLC  
  — QOL Medical is the manufacturer of a treatment to relieve the symptoms of Genetic Sucrase-Isomaltase Deficiency (GSID)

Outline

#1  Quick carbohydrate overview – as it relates to Genetic Sucrase-Isomaltase Deficiency (GSID)
#2  Why should I test for GSID? It’s so rare.
  - Prevalence
#3  Who should be screened for GSID?
  - Identifying the Patient
#4  What kind of diagnostic testing is available?
  - Diagnostic Tools
#5  How do I treat GSID?
  - Treatment Options
Classification of Carbohydrates

Genetic Sucrase-Isomaltase Deficiency (GSD) Activities

- **Sucrase**
  - Sucrose = fructose + glucose
  - Hydrolyzes α-1,2- and α-1,4-glucosidic bonds
  - Digests sucrose, maltose, and maltotriose

- **Isomaltase**
  - Maltose = glucose + glucose
  - Hydrolyzes α-1,6 linkages
  - Digests isomaltose, maltose, maltotriose, and limit dextrins

- GSD and maltase-glucoamylase (MGAM) are intricately involved in sucrose and starch digestion²,³

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Carbohydrate Digestion

Sucrose Digestion and Possible Clinical Implications

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information on slides 37-39 and Full Prescribing Information passed out at the beginning of this presentation. Sucraid® may cause a serious allergic reaction.
Starch Digestion and Possible Clinical Implications

- Starch → Dextrins → Maltose → Glucose + Glucose
- **Colon Fermentation**
  - Pulls in excess water
  - +/- Low Energy
  - +/- Low Blood Sugar
  - Gas, Disention
  - Bloating, +/- Diarrhea

#1 Why Should I Test for GSID? It's So Rare

Prevalence

Let's Talk Briefly about Prevalence...

- Historical data (1960 – 1970) for Genetic Sucrase-Isomaltase Deficiency (a genetically inherited autosomal recessive disorder)
  - 5 – 10% in Greenland Eskimos
  - 3 – 7% in Canadian native peoples
  - 3% in Alaskans of native ancestry
  - <0.2% in non-Hispanic whites
- But, look at what we have learned since then...

Genetic Sucrase-Isomaltase Deficiency (GSID)

- GSID originally thought to cause symptoms in autosomal recessive (homozygous) inheritance only
- Recent studies have demonstrated carriers may also present with symptoms

References:
Potential Etiologies of Sucrase Deficiency

Genetic Sucrase-Isomaltase Deficiency (GSID) Genotypes
- Congenital Sucrase-Isomaltase Deficiency (CSID) (Compound Heterozygotes)
- GSID Symptomatic Carriers (Simple Heterozygotes)

Secondary Sucrase Deficiency
- Celiac Disease
- Bacterial Overgrowth
- Crohn’s Disease
- Other (e.g., blunted villi)

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information on slides 37-39 and Full Prescribing Information passed out at the beginning of this presentation. Sucraid® may cause a serious allergic reaction.

GSID Genetic Prevalence Study
Genetic Results by Primary Symptom

<table>
<thead>
<tr>
<th>Primary Variants (Heterozygotes)</th>
<th>Abdominal Pain</th>
<th>Diarrhea</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>n= 375</td>
<td>n= 375</td>
</tr>
<tr>
<td>Primary Variants (Heterozygotes)</td>
<td>13</td>
<td>8</td>
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<tr>
<td>Secondary Variants (Heterozygotes)</td>
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<td>2</td>
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<tr>
<td>Compound Variants (Heterozygotes)</td>
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<tr>
<td>Total</td>
<td>16</td>
<td>10</td>
</tr>
<tr>
<td>Rate</td>
<td>4.3%</td>
<td>2.7%</td>
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</table>

Frequency of Sucrase Deficiency in Mucosal Biopsies Review of Disaccharidase Analyses
N=27,375

- Total Sucrase Deficiency (1.2 SD< Norm) 9.3%
- All Other Results

Prospective Disaccharidase Testing in Children with Recurrent Abdominal Pain

<table>
<thead>
<tr>
<th>Pilot Study N=28</th>
<th>% of total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low Lactase</td>
<td>18</td>
</tr>
<tr>
<td>Low Sucrase</td>
<td>4</td>
</tr>
<tr>
<td>Low Maltase</td>
<td>5</td>
</tr>
<tr>
<td>Low Glucoamylase</td>
<td>4</td>
</tr>
<tr>
<td>Pan-Disaccharidase Deficiency (all 4 low)</td>
<td>3</td>
</tr>
</tbody>
</table>

Retrospective Study of 963 Symptomatic Children Undergoing Upper Endoscopy and Disaccharidase Testing

**PRIMARY RESULTS**

5,362 EGDs

- 903 (18%) Disaccharidase Testing
  - 430 (46%) LD only
  - 73 (7.6%) SD
  - 44 (60%) Pan-Diastase
  - 25 (34%) With LD
  - 4 (6%) Isolated

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#2 Who Should Be Screened for GSD?

Identifying the Patient

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Checklist for Considering GSD

- **Reported Symptoms:**
  - Chronic diarrhea
  - Abdominal pain
  - Abdominal distension
  - Excess gas
  - Excoriated buttocks
  - Vomiting
  - Failure to thrive
  - Weight loss
  - Constipation
  - Acid reflux
  - Burping

- **Possible Diagnosis:**
  - Viral/bacterial gastroenteritis
  - Parasitic diarrhea
  - Lactose intolerance
  - Celiac Disease
  - Inflammatory Bowel Disease
  - Crohn’s Disease
  - Ulcerative Colitis
  - Cystic Fibrosis
  - Irritable Bowel Syndrome
  - **Genetic Sucrase-Isomaltase Deficiency**

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Initial Presentation of GSD in Infancy

- May not present until ~ 6 months - 1 year
  - Breast milk and standard infant formula (CHO source is lactose) are tolerated
  - First illness; antibiotic (high in sucrose)
  - Introduction of sucrose and/or starch
  - Formula, cereals, fruits, vegetables, and/or juice

- Often misdiagnosed as allergy or intolerance to cow’s milk or soy protein
  - Changing baby formula may actually worsen symptoms and delay diagnosis

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Think about the **Toddler with Chronic Diarrhea**...

- Started with diarrhea around 6 months of age
- Hospitalized with vomiting and diarrhea
- Susceptible to infection, URIs
- Allergy testing all negative
- Tried Alimentum®, EleCare®, Neocate® Splash, Peptamen Junior®

Think about the **Toddler with Chronic Diarrhea**...

- Reports no sweets and no nuggets or fries from McDonald’s
- Typical intake at 3 y/o
  - B: ham, egg
  - Sn: potato chips or corn chips
  - L: eats meat off sandwich
  - D: mac and cheese
  - Drinks: Peptamen Junior®
- Endoscopy with small bowel biopsies for disaccharidase assay
  - Low-sucrase
  - Low-glucoamylase (maltase)
  - Normal lactase
  - Normal mucosa

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Think about Your **Older Patients** Diagnosed with IBS - Could It Be GSID?

- You’ve tried medications and various diets
- Patient finds some relief with Low FODMAP® diet
  - But still has lingering symptoms: diarrhea, abdominal pain, gas, and bloating
  - And you believe they are compliant with the diet
- Consider testing for GSID (see #4 for diagnostic options)
- Consider a trial of SucraLac® (sacrosidase) Oral Solution therapy and diet modification (see #5 for treatment options)

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#3 What Kind of Diagnostic Testing Is Available?

**Diagnostic Tools**

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Please see SucraLac® Important Safety Information on slides 37-39 and Full Prescribing Information passed out at the beginning of this presentation. SucraLac® may cause a serious allergic reaction.
Diagnostic Options for GSID

- **Diagnostic Tests**
  - Upper Endoscopy - Small Bowel Biopsy - Disaccharidase Assay
    - “Gold Standard”
  - Trial Prescription of Sucraid® (sacrosidase) Oral Solution
    - Prescription/order forms on Sucraid.net website

- **Tests That Aid in Diagnosis**
  - Sucrose Hydrogen Methane Breath Test (SHMBT)
    - QOL Medical offers this test free of charge through Commonwealth Labs
  - Genetic Testing (buccal swab or saliva)
    - Genetic Test Kits can be ordered free of charge from Sucraid.net

"Gold Standard" Diagnostic Test:
Disaccharidase Activity Assay

<table>
<thead>
<tr>
<th>Normal Range (µmol glucose/min/g protein)</th>
<th>Deficient (µmol glucose/min/g protein)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sucrase</td>
<td>29.0 - 79.8</td>
</tr>
<tr>
<td>Maltase</td>
<td>98.0 - 223.6</td>
</tr>
<tr>
<td>Palatinase</td>
<td>4.6 - 17.6</td>
</tr>
<tr>
<td>Lactase</td>
<td>16.5 - 32.5</td>
</tr>
</tbody>
</table>

Pan-Disaccharidase Deficiency

- Do not discredit a Disaccharidase Assay where all 4 enzymes are deficient (pan-disaccharidase deficiency)
- In general, as people age, they are more likely to have a lactase deficiency:
  - Congenital lactase deficiency (in infants) is rare
  - Approximately 65 percent of the human population has a reduced ability to digest lactose after infancy
  - It makes sense that an older child and certainly an adult could have both a lactase deficiency and Genetic Sucrase-Isomaltase Deficiency
  - QOL Observational Study of 49 Sucraid® patients: 25% also had a lactose intolerance
- Treat for both GSID and lactose intolerance

Sucraid® (sacrosidase) Oral Solution Therapy

- If other more common GI disorders have been ruled out, it may be warranted to prescribe Sucraid® as a one to two week trial to see if the patient responds.
- Order/prescription forms are available at www.sucraid.net
- Please see Sucraid® Important Safety Information and details on accessing the Full Prescribing Information in slides 37-39
 Sucrose Hydrogen Methane Breath Test (SHMBT)

- A noninvasive, functional test to aid in identifying and diagnosing patients suffering with Genetic Sucrase-Isomaltase Deficiency (GSID)
- SHMBT kits are available to healthcare professionals, including dietitians, for patient distribution at no charge (through QOL Medical, LLC)
- Test results will be sent to the physician’s office within 48 hours of receiving the completed test kit

 Prior to the SHMBT, patients are instructed to discontinue certain medications (probiotics, antibiotics) and follow a low-carbohydrate diet one day before the test
- After a 12-hour overnight fast, the patient starts with a baseline breath sample, drinks a sucrose solution, and completes a series of breath samples (test is ~3 hours in all)
  - Patients with GSID may experience GI symptoms due to the consumption of the sucrose solution and may prefer to take the test at home
  - Both hydrogen and methane gas production are measured
    - Mal-digested sucrose results in an increase of these gases, due to bacterial fermentation

Genetic Testing

- Genetic Testing – no charge for the cost of the kit or testing
- Order genetic test kit from www.sucraid.net
- Buccal swab
- Test kit sent out to LabCorp
- Detects 37 known mutations
- Can help rule GSID in, but may not necessarily rule it out

#4 How Do I Treat GSID?

Treatment Options
Patient Identified! What Are the Treatment Options?

- Diet alone (low-sucrose and low-starch)
- Sucraid® (sacrosidase) Oral Solution - enzyme replacement therapy for the treatment of the genetically determined sucrase deficiency which is part of congenital sucrase-isomaltase deficiency (CSID)
  - Although Sucraid® provides replacement therapy for the deficient sucrase, it does not provide specific replacement therapy for the deficient isomaltase
- Combined diet and Sucraid® therapy

Infant Diet Guidelines for Patients Diagnosed with GSD

- Formula
  - Breast milk
  - Standard infant formula (lactose-based)
  - Abbott Nutrition RCF® no added carbohydrate soy-based formula
  - Add dextrose or fructose
  - Add water
  - Provide mixing instructions

Available on: CSIDcares.org Sucraidassist.com Sucraid.net

Starting Solids

- Start solids as soon as age appropriate; don’t delay
- Start Sucraid® (sacrosidase) Oral Solution therapy when starting solids
  - Sucraid® has been tested in clinical trials with children 5 months of age and older
- Start with low-sucrose, low-starch, pureed vegetables, fruits, and then meats; do not start with cereals
**INDICATION:**
- Sucraid® (sacrosidase) Oral Solution is an enzyme replacement therapy for the treatment of the genetically determined sucrase deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID).

**IMPORTANT SAFETY INFORMATION:**
- Although Sucraid® provides replacement therapy for the deficient sucrase, it does not provide specific replacement therapy for the deficient isomaltase.
- Do not use Sucraid® (sacrosidase) Oral Solution with patients known to be hypersensitive to yeast, yeast products, papain, or glycerin (glycerol).

**IMPORTANT SAFETY INFORMATION (cont'd.):**
- Sucraid® may cause a serious allergic reaction. Patients should stop taking Sucraid® and get emergency help immediately if any of the following side effects occur: difficulty breathing, wheezing, or swelling of the face. Care should be taken when administering initial doses of Sucraid® to observe any signs of acute hypersensitivity reaction.
- Adverse reactions as a result of taking Sucraid® may include worse abdominal pain, vomiting, nausea, diarrhea, constipation, difficulty sleeping, headache, nervousness, and dehydration.
- Before prescribing Sucraid® to diabetic patients, the physician should consider that Sucraid® will enable sucrose hydrolysis and the absorption of those hydrolysis products, glucose and fructose.

**IMPORTANT SAFETY INFORMATION (cont'd.):**
- The effects of Sucraid® have not been evaluated in patients with secondary (acquired) disaccharidase deficiency.
- DO NOT HEAT SOLUTIONS CONTAINING SUCRAID®. Do not put Sucraid® in warm or hot fluids. Do not reconstitute or consume Sucraid® with fruit juice since the acidity of the juice may reduce the enzyme activity of Sucraid®. Half of the reconstituted Sucraid® should be taken at the beginning of the meal or snack and the other half during the meal or snack.
- Sucraid® should be refrigerated at 36°F-46°F (2°C-8°C) and should be protected from heat and light.
- FULL PRESCRIBING INFORMATION was provided at the beginning of this presentation and can be accessed online at sucraid.net/pi.pdf.

**Dosing and Administration:**
- Sucraid® must be kept refrigerated
- Dosing:
  - 1 mL (28 drops) if ≤ 15 kg (≤ 33 lbs)
  - 2 mL (56 drops) if > 15 kg (> 33 lbs)
- Mixing:
  - Mix Sucraid® in 2 – 4 ounces of milk, water, or sucrose-free, starch-free formula.
  - Do not mix in hot beverages; do not heat after mixing.
  - Administer Sucraid® with every meal or snack.
  - Take ¼ at beginning of meal/snack and other ¼ during meal/snack.
Treatment Plan for Previously Discussed Toddler (GSID Confirmed by Biopsy)

- Start Sucraid® (sacrosidase) Oral Solution therapy
- Modified diet/elimination (sucrose and starch)
- Consider changing formula to RCF® (Abbott) or KetoCal® (Nutricia)

A Comparison of Other Elemental Formulas Often Tried, But Failed...

<table>
<thead>
<tr>
<th>FORMULA</th>
<th>PROTEIN SOURCE</th>
<th>CARB SOURCE</th>
<th>FAT SOURCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Similac Alimentum® (Abbott)</td>
<td>casein hydrolysate</td>
<td>sugar, modified tapioca starch</td>
<td>safflower oil, MCT, soy oil</td>
</tr>
<tr>
<td>Be-Care® (Abbott)</td>
<td>amino acids</td>
<td>corn syrup solids</td>
<td>safflower oil, MCT, soy oil</td>
</tr>
<tr>
<td>Neocate® Jr® (Nutricia)</td>
<td>amino acids</td>
<td>corn syrup solids, vegetable oil, canola oil, soy oil</td>
<td>vegetable oil, canola oil, soy oil</td>
</tr>
<tr>
<td>Splash® (Nutricia)</td>
<td>amino acid, maltodextrin, sugar</td>
<td>corn syrup solids, vegetable oil, canola oil, soy oil</td>
<td>vegetable oil, canola oil, soy oil</td>
</tr>
<tr>
<td>Peptamen® Jr® (Nestle)</td>
<td>hydrolyzed whey protein</td>
<td>maltodextrin, sugar, cornstarch</td>
<td>MCT, soybean oil, canola oil</td>
</tr>
</tbody>
</table>

Formula Options for Our Toddler...

<table>
<thead>
<tr>
<th>FORMULA</th>
<th>PROTEIN SOURCE</th>
<th>CARB SOURCE</th>
<th>FAT SOURCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Portagen® (Mead Johnson)</td>
<td>sodium caseinate</td>
<td>corn syrup solids, sugar</td>
<td>MCT, corn oil</td>
</tr>
<tr>
<td>RCF® (Abbott)</td>
<td>soy protein isolate</td>
<td>none</td>
<td>safflower oil, soy oil, coconut oil</td>
</tr>
<tr>
<td>KetoCal® 4:1 (Nutricia)</td>
<td>milk protein (casein, whey)</td>
<td>corn syrup solids</td>
<td>vegetable oil</td>
</tr>
</tbody>
</table>

Child or Adult Diet Guidelines for Patients Diagnosed with GSID

- Begin Sucraid® (sacrosidase) Oral Solution therapy
- Elimination Diet (2 weeks) – eliminate the culprits
  — Sucrose
  — Starch, maltose
  — Others
- Symptom improvement is generally seen within these first two weeks
- Induction Diet – establish tolerance to sucrose and starch
  — Sucrose (Sucraid® may allow for a nearly “normal” intake of sucrose)
  — Starch tolerance will vary significantly (Sucraid® does not replace isomaltase)

Please see Sucraid® Important Safety Information on slides 37-39 and Full Prescribing Information passed out at the beginning of this presentation. Sucraid® may cause a serious allergic reaction.
Why Do Patients with GSID Need to See a Registered Dietitian?

- Sucrose and starch modifications are difficult to understand, making the diet hard to follow for newly diagnosed patients:
  - Food labels do not provide sucrose and starch content
  - Most nutrient databases do not have complete sucrose and starch data
  - People do not think in terms of sucrose and starch; they are more likely to think in terms of sugars and carbohydrates, but even those terms are not always familiar to the general population

Why Do Patients with SI Deficiency Need to See a Registered Dietitian?

- Continued…
  - Many patients may have other diagnoses and diet restrictions to contend with:
    - Lactose intolerance
    - Wheat or gluten sensitivity, Celiac Disease, or food allergies
    - IBS/low FODMAPs diet
    - Milk protein intolerance
  - They may be given a list of what NOT to eat, but do not know what they CAN eat!
  - Improper diet modifications and restrictions increase risk of nutrient deficiencies
Nutrition Topics to Cover with Patients

- What is sucrose?
- What is starch?
- What are acceptable alternatives to cow’s milk?
- How much sucrose/starch is too much?
- How much sucrose/starch is not enough?
- How to read a food label?
- What about lactose, fructose?
- What artificial sweeteners are acceptable?
- Is gluten-free low in starch?
- What about 100% fruit juice?
- What about FODMAPS, SCD?
- How do I meet calorie needs?
- Where do I buy fructose and dextrose?

Grams of Sucrose per Meal/per Day?

- Compliance with a sucrose-free diet is very difficult
- Following an elimination diet, sucrose can be increased by ~0.5 g/day or more
- Add back sucrose via fruits and healthier “sugar” foods first
- Sucraid® (sacrosidase) Oral Solution will cover a significant amount of sucrose in the diet, but tolerance is variable and depends on the individual

Grams of Starch per Meal/per Day?

- Although Sucraid® (sacrosidase) Oral Solution provides replacement therapy for the deficient sucrase, it does not provide specific replacement therapy for the deficient isomaltase; starch tolerance is highly variable
- “Cracker Test”: adding 1 Saltine cracker/day (2 g starch per day)
- Ann McMeans, RD suggests 120 grams/day may be tolerated
- The type of starch may allow more grams per day
- Chewing food well – exposing to amylase longer
- Some take over-the-counter “starch enzymes”

Nutrient Data for Sucrose and Starch

- Sucrose and starch information are not found on food labels
- Sucrose and starch data are not a part of most nutrient databases; USDA has limited information, but adding to it
- University of Minnesota Nutrition Data System for Research (NDSR) has complete sucrose and starch data
- Total carbohydrates, total sugars, and fiber are on Nutrition Facts Label

Total CHO’s = total sugars – fiber = starch

- Type(s) of sugar and starch sources are listed on the ingredient label, but patients must learn to be ingredient detectives
QOL Observational Study

- 49 subjects on Sucraid® (sacrosidase) Oral Solution
  - >3 months on therapy
  - Age ranged 1 – 45 years
- 24-hour dietary recalls; three consecutive days

<table>
<thead>
<tr>
<th>% of Total Kcals</th>
<th>AVG for All Subjects (%)</th>
<th>DRI (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CHO</td>
<td>42</td>
<td>45-65</td>
</tr>
<tr>
<td>FAT</td>
<td>39</td>
<td>25-40</td>
</tr>
<tr>
<td>PRO</td>
<td>18</td>
<td>5-35</td>
</tr>
</tbody>
</table>

Please see Sucraid® Important Safety Information on slides 37-39 and Full Prescribing Information passed out at the beginning of this presentation. Sucraid® may cause a serious allergic reaction.


Tolerance to Sweeteners

- Sweeteners “tolerated by most”
  - Dextrose and Fructose
  - High Fructose Corn Syrup
  - Aspartame (NutraSweet®)

- Sweeteners “tolerated by some”
  - Agave nectar (primarily glucose and fructose)
  - Equal® (aspartame based)
  - Honey (primarily glucose and fructose)
  - Saccharin (Sweet’N Low®)
  - Stevia®
  - Sucralose (Splenda®)
  - Sugar alcohols

- Top Five Sucrose Sources:
  - cake/pie, donut, ice cream, frozen desserts, espresso/chocolate milk
- Top Five Starch Sources:
  - pizza, pasta, potato, brown rice, bread
- Top Five Maltose Sources:
  - breakfast cereal, chips, pancake syrup, tortilla, CLIF bar
  - Increased maltose associated with increased GI symptoms
Tolerance to Sweeteners

- Sweeteners to "Avoid"; Sucrose:
  - Beetsugar
  - Brown sugar
  - Cane juice, cane sugar
  - Caramel
  - Coconut sugar
  - Confectioner’s sugar
  - Date sugar
  - Maple syrup
  - Molasses
  - Raw sugar
  - Sorbitol
  - Sugar

Starch Ingredients That May Not Be Tolerated

- Limit dextrins
- Maltodextrin
- Modified tapioca starch
- Glucose polymers
- Maltose
  - Brown rice syrup
  - Corn syrup solids
  - Malt

Fruit Juice

<table>
<thead>
<tr>
<th>Fruit Juice</th>
<th>Sucrose</th>
<th>Starch</th>
<th>Glycemic Index</th>
<th>Fructose</th>
<th>Glucose</th>
<th>Maltose</th>
<th>Lactose</th>
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<tbody>
<tr>
<td>Apple</td>
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<td>0</td>
<td>39</td>
<td>7.1</td>
<td>2.3</td>
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<tr>
<td>Carrot</td>
<td>0.9</td>
<td>0</td>
<td>39</td>
<td>6.8</td>
<td>0</td>
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<td>Melons</td>
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<td>Prunes</td>
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<td>Apple-Grape Drink</td>
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<td>4.6</td>
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Differences Between Elimination Diets

<table>
<thead>
<tr>
<th></th>
<th>Low FODMAPs</th>
<th>Low Sucrose, Low Starch</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dairy</td>
<td>No (some cheese)</td>
<td>Yes</td>
</tr>
<tr>
<td>Fat</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Protein (meats)</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Starch/grains</td>
<td>Yes (selected)</td>
<td>No</td>
</tr>
<tr>
<td>Fruits</td>
<td>Yes (low fructose)</td>
<td>Yes (low sucrose)</td>
</tr>
<tr>
<td>Vegetables</td>
<td>Yes (selected)</td>
<td>Yes (low starch)</td>
</tr>
<tr>
<td>&quot;Sugars&quot;</td>
<td>No fructose;</td>
<td>No sucrose;</td>
</tr>
<tr>
<td></td>
<td>Yes (sugar, syrup, jelly)</td>
<td>Yes (fructose, dextrose)</td>
</tr>
</tbody>
</table>
Case Study - RD Intervention

- 4-year-old female presented to RD with IBS for education on low FODMAPs diet
- Patient showed some improvement on diet, but still with GI symptoms
  - FODMAPs does not restrict sucrose and starch
- RD recalled our conversation about SI deficiency and Sucraid® (sacrosidase) Oral Solution therapy
- RD probed and thought perhaps this could be “one of those patients”
- RD recommended the MD order a trial of Sucraid®
- After 10 days on Sucraid®, BMs normalized and all other symptoms improved
  - This child’s symptoms began around 1 year of age
  - She was 4 years old before GI symptoms resolved
  - Genetic testing was negative

Please see Sucraid® Important Safety Information on slides 37-39 and Full Prescribing Information passed out at the beginning of this presentation. Sucraid® may cause a serious allergic reaction.

Allowed or Not Allowed on Elimination Phase of A Low-Sucrose, Low-Starch Diet

- **Allowed on Elimination Phase of**
  - Apples
  - Olives
  - Bananas
  - Crackers
  - Cottage cheese
  - Blueberries
  - Gluten-free
  - Yogurt
- **Not Tolerated by most**
  - Nuts
  - Grapes
  - Butter
  - Green beans

QOL Medical, LLC Offers a Variety of Services to Healthcare Providers and Their Patients

- **One Patient Services - SucraidASSIST™**
  - Nurse Case Manager
  - Point of contact for all needs (patients and clinicians)
  - Prescriptions, forms, paperwork, tracking
  - All questions
  - Insurance Specialist
  - Dietitian
  - Peer Coach

- **Websites:**
  - SucraidAssist.com
  - CSIDcares.org
  - Sucraid.net

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Thank You!

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Questions?

Thank you for the opportunity to share this presentation with you today!