Genetic Sucrase-Isomaltase Deficiency (GSID)

...what you might be missing

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QOL Medical, LLC
Learning Objectives

The attendee will gain a basic understanding of Genetic Sucrase-Isomaltase Deficiency (GSID) and be able to...

a. Identify patients for further screening to assess for a possible diagnosis of GSID

b. Make recommendations to the patient's physician regarding further diagnostic testing for GSID

c. Provide appropriate and effective nutrition counseling, education, and treatment plans for patients diagnosed with GSID
Disclosures

- Employed by QOL Medical, LLC, the maker of Sucraid® (sacrosidase) Oral Solution, the enzyme replacement therapy for the treatment of genetically determined sucrase deficiency, which is a 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

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Review of the Literature

- *Journal of Pediatric Gastroenterology and Nutrition (JPGN)*; November 2012; Volume 55; Supplement 2.

- Research progress reported at the 50th anniversary of the discovery of Congenital Sucrase-Isomaltase Deficiency (CSID) Workshop. Guest Editor: Buford L. Nichols Jr, MD, MS

Congenital Sucrase-Isomaltase Deficiency (CSID) was first described by Weijers and colleagues in 1960\(^1\). CSID was described as a rare, autosomal recessive, inborn error of metabolism\(^2\). Mutations in the SI gene prevent normal synthesis and transport of the protein responsible for sucrase and isomaltase\(^2\). CSID results in the maldigestion of sucrose and starch, resulting in significant GI symptoms. Historical prevalence data:

- 5 - 10% in Greenland Eskimos
- 3 - 7% in Canadian native peoples
- 3% in Alaskans of native ancestry
- <0.2% in non-Hispanic whites

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Historical Perspective
Congenital Sucrase-Isomaltase Deficiency (CSID)

- Early studies with Baker’s yeast (*Saccharomyces cerevisiae*), for the treatment of CSID in the 1980s

- Sucraid® (*sacrosidase*) Oral Solution trials began in the early ’90s by Dr. Wil Treem, Duke Pediatrics, GI

- Sucraid® was FDA approved in 1998

- Tremendous strides have been made over the last 50 years to better understand, diagnose, and treat CSID
  — Most recently in the genetics and prevalence of CSID

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Please see Sucraid® (*sacrosidase*) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
The Genetics of Sucrase-Isomaltase (SI) Deficiency$^{1,2}$

- May be more prevalent than previously thought...

## CSIDgps Study
### Genetic Results by Primary Symptom

<table>
<thead>
<tr>
<th></th>
<th>Diarrhea</th>
<th>Abdominal Pain</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>n</td>
<td>375</td>
<td>375</td>
<td>750</td>
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<tr>
<td>Primary Variants</td>
<td></td>
<td></td>
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<tr>
<td>(Heterozygotes)</td>
<td>15</td>
<td>8</td>
<td>23</td>
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<tr>
<td>Secondary Variants</td>
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<td></td>
<td></td>
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<tr>
<td>(Heterozygotes)</td>
<td>0</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Compound Heterozygotes</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>16</td>
<td>10</td>
<td>26</td>
</tr>
<tr>
<td><strong>Rate</strong></td>
<td><strong>4.3%</strong></td>
<td><strong>2.7%</strong></td>
<td><strong>3.5%</strong></td>
</tr>
</tbody>
</table>
Potential Etiologies of Sucrase Deficiency

Genetic Sucrase-Isomaltase Deficiency (GSID) Genotypes

- Classic CSID
  - (Compound Heterozygotes)

- GSID
  - Symptomatic Carriers
    - (Simple Heterozygotes)

Secondary Sucrase Deficiency

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

Sucraíld® (sacrosidase) Oral Solution is indicated for the treatment of the genetically determined sucrase deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID).

- Although Sucraíld® provides replacement therapy for the deficient sucrase, it does not provide specific replacement therapy for the deficient isomaltase.

Sucraíld® may cause a serious allergic reaction.

Please see Sucraíld® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32.
Frequency of Sucrase Deficiency in Mucosal Biopsies Review of Disaccharidase Analyses

Study of Idiopathic Sucrase Deficiency
N= 27,875

- Sucrase Deficient (1.2 SD< Norm)
- Normal

Carbohydrate Digestion
Carbohydrate Digestion

**Sucrase-Isomaltase Complex** is composed of 2 subunits:

1. **Sucrase** digests:
   - Sucrose  $\rightarrow$ Glucose and Fructose
   - Maltose  $\rightarrow$ Glucose and Glucose ($\alpha$-1,4)
   - Maltotriose  $\rightarrow$ Glucose, Glucose, and Glucose ($\alpha$-1,4)

2. **Isomaltase** digests:
   - Isomaltose  $\rightarrow$ Glucose and Glucose ($\alpha$-1,6)
   - Maltose  $\rightarrow$ Glucose and Glucose ($\alpha$-1,4)
   - Maltotriose  $\rightarrow$ Glucose, Glucose, and Glucose ($\alpha$-1,4)
   - Limit dextrins  $\rightarrow$ Glucose ($\alpha$-1,4 and $\alpha$-1,6)
Carbohydrate Digestion

Sucrose Digestion and Possible Clinical Implications

Sucrose → COLON Fermentation

- Pulls in excess water
- DIARRHEA

Sucrose → COLON Fermentation

- Pulls in excess water
- DIARRHEA

Sucrose → SUCRASE

Glucose + Fructose

- LOW ENERGY
- LOW BLOOD SUGAR

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Starch Digestion and Possible Clinical Implications

Starch → dextrins → Maltose → Glucose + Glucose

- Amylase - Alpha glucoamylase - Amylase -- Isomaltase

Colon

Fermentation

Pulls in excess water

GAS, DISTENTION

BLOATING, +/- DIARRHEA

TIME

+- LOW ENERGY

+- LOW BLOOD SUGAR
Diagnostic Options for SI Deficiency

- **Upper Endoscopy – Small Bowel Biopsy – Disaccharidase Assay**
  - “Gold Standard”

- **Therapeutic Response Dose of Sucraïd® (sacrosidase) Oral Solution**
  - Prescription/order forms on Sucraïd.net website

- **Tests That Aid in Diagnosis**
  - Sucrose Intolerance Hydrogen Breath Test (SIHBT)
  - SI Genetic Testing (buccal swab or saliva)

Please see Sucraïd® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraïd® may cause a serious allergic reaction.
“Gold Standard” Diagnostic Test: Disaccharidase Activity Assay

- **Disaccharidase Levels:**

<table>
<thead>
<tr>
<th>ENZYME</th>
<th>Normal Range uM/min/g protein</th>
<th>Abnormal Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lactase</td>
<td>16.5 – 32.5</td>
<td>&lt; 15</td>
</tr>
<tr>
<td>Sucrase</td>
<td>25.0 – 79.8</td>
<td>&lt; 25.0</td>
</tr>
<tr>
<td>Maltase</td>
<td>100.0 – 223.6</td>
<td>&lt; 100.0</td>
</tr>
<tr>
<td>Palatinase</td>
<td>5.0 – 17.6</td>
<td>&lt; 5.0</td>
</tr>
</tbody>
</table>

- **Classic CSID:** absent or low sucrase, low maltase and palatinase, normal lactase; mucosal histology is normal

Variable Use of Disaccharidase Assays Among Pediatric Gastroenterologists Evaluating Abdominal Pain

The analyses compared the frequency of EGD, the number of endoscopic records from patients who underwent EGD and the number of disaccharidase assays performed (Tested) and the number of disaccharidase biopsies performed and assay results.

Recent work suggests the frequency of lactase deficiency (LD) is 42% and is very high upon disaccharidase testing at the time of upper endoscopy (Nichols et al., 2012).

The compound heterozygosity of sucrase deficiency is becoming better understood.

Among patients undergoing EGD (N=3,344), 73% (7.6%) SD and LD only (44% (60%) LD only; 25% (34%) With LD; 4% (5%) isolated LD.

There is a need for further prospective studies on the prevalence and significance of disaccharidase deficiencies and the best diagnostic procedures to establish the diagnosis.

REGRESSION ANALYSIS

TOTAL DISACCHARIDASES VS SUCRASE DEFICIENT

Abdominal Pain vs Sucrase Deficient

Diarrhea vs Sucrase Deficient

Total Disaccharidases vs Lactase Deficient

Abdominal Pain vs Lactase Deficient

Diarrhea vs Lactase Deficient

DISACCHARIDASE TEST RESULTS

CONCLUSION

The analyses compared the frequency of EGD, the number of disaccharidase biopsies performed and assay results.

Among patients undergoing EGD (N=3,362), disaccharidase testing was performed in 963 (18%). The primary indication for performing EGD was abdominal pain (N=3,344).

Among the physicians who performed an EGD in patients with abdominal pain, the percent of disaccharidase assays performed varied widely among physicians, ranging from 1.6% to 64.5%.

Regression analyses revealed significant correlations between the number of EGD performed (N) and the number of patients found with SD (p=0.019) and LD patients (p=0.003) and significant correlations between the numbers of disaccharidase assays performed (Tested) and the number of SD and LD patients (each, p<0.0001).

Cohen, Stanley A., MD1; Oloyede, Hannah, MPH2

1. Children's Center for Digestive Health Care
2. Children's Healthcare of Atlanta
Atlanta, Georgia
Variable Use of Disaccharidase Assays Among Pediatric Gastroenterologists Evaluating Abdominal Pain

**PRIMARY RESULTS**

5,362 EGDs

963 (18%) Disaccharidase Testing

430 (45%) LD only

73 (7.6%) SD

44 (60%) Pan-Disaccharidase

25 (34%) With LD

4 (5%) Isolated
Sucrose Intolerance Hydrogen Breath Test (SIHBT)

- A noninvasive test for older children and adults

- SIHBT test kits are available to healthcare professionals for patient distribution at no charge (via QOL Medical)
  - Test kits may be ordered by calling 1-800-705-1962
  - Tests may be done in the physician’s office or at the patient’s home
  - There is some preparation prior to the breath test (diet and medication restrictions; fasting)
  - Patients must drink 50 grams of sucrose (25 grams for children). Patients with sucrase deficiency may experience symptoms due to the large consumption of sucrose

- Test results are sent to the physician’s office within 24 hours of receiving the completed breath test kit
Genetic Testing

- Genetic Testing – no charge for the cost of the kit or testing
- Buccal swab
- >37 known mutations
- Order genetic test kit from [www.sucraid.net](http://www.sucraid.net); send out to LabCorp
- A positive genetic test supports the diagnosis of GSID. The test evaluates for the presence of 37 known pathogenic variants. A negative test result demonstrates the absence of one or more of these variants. However, other unidentified pathogenic variants may exist, so a negative test is not conclusive for absence of the disease.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
If the diagnosis is doubtful and other more common GI disorders have been ruled out, it may be warranted to conduct a short therapeutic trial (e.g., one week) with Sucraid® to assess response in a patient suspected of sucrase deficiency.

Visit [www.sucraid.net](http://www.sucraid.net) for enrollment forms.

Please see Sucraid® Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32.
What Might a GSID Patient Look Like?

- **Common Symptoms:**
  - Chronic diarrhea*
  - Abdominal pain
  - Abdominal distension
  - Gas
  - Excoriated buttocks
  - Failure to thrive
  - Weight loss

- **Other symptoms:**
  - Vomiting
  - Constipation

- **Possible Diagnoses:**
  - Viral/bacterial gastroenteritis
  - Parasitic diarrhea
  - Lactose intolerance
  - Fructose intolerance
  - Milk protein intolerance
  - Celiac Disease
  - Inflammatory Bowel Disease
  - Crohn’s Disease
  - Ulcerative Colitis
  - Cystic Fibrosis
  - Small intestinal bacterial overgrowth (SIBO)
  - Irritable Bowel Syndrome
  - **Genetic Sucrase-Isomaltase Deficiency**
Initial Presentation of CSID 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

Infant Diet Guidelines for Patients Diagnosed With GSID

(Infant Diet Guide)
Guidelines for Infants with Genetic Sucrose-Isomaltase Deficiency (GSID)

Starting Sucraid® (sacrosidase) Oral Solution Therapy with a Low-Sucrose, Low-Starch Diet

Key Points:
- Start with breast milk or sucrose-free, starch-free infant formula.
- Add Sucraid® when introducing solid foods (5-6 months).
- Consult with a Registered Dietitian (RD) for diet assessment, education, and follow-up.

Note: These diet recommendations are for general guidance only. Every infant is different, and the diet should be individualized. The dietitian may develop a diet plan in consultation with a gastroenterologist and geneticist.

Birth - 4 months:
- Breast milk or sucrose-free, starch-free infant formula (Enfamil® Premium, Similac® Alimentum) at the infant's usual needs for the first 4 months of life.

4 - 6 months:
- The American Academy of Pediatrics (AAP) recommends starting solid foods around 6 months of age. Consider a dietitian to determine developmental readiness for starting solid foods.
- Adding Sucraid® with meals and snacks allows increased variety in the diet.
- Do not start with baby cereal; it is high in starch and may cause symptoms of malabsorption.
- Start with puréed vegetables low in sucrose and starch (see food lists at CIBrox.com).
- May add puréed fruits low in sucrose and starch (see food lists at CIBrox.com).
- May add cooked meats (good source of iron).

6 - 8 months:
- May be ready to switch from purée.
- May advance to mashed vegetables low in sucrose and starch (see food lists at CIBrox.com).
- May advance to mashed fruits low in sucrose and starch (see food lists at CIBrox.com).
- May advance to very soft, plain mashed meats (no breadcrumb).

8 - 10 months:
- May add plums, unsweetened yogurt, cottage cheese, or part-skimmed cheese.
- May add eggs (uncooked or hard boiled and cut into small pieces) for safe chewing and swallow.

10 - 12 months:
- May be ready for low sucrose, low-starch, solid foods (processed or cut into small pieces for safe chewing and swallow).

Over 12 months:
- May introduce whole eggs.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
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
Purchasing Fructose and Dextrose

- Some grocery stores may sell fructose and/or dextrose
- Some bakeries or breweries may also sell dextrose
- Fructose and dextrose can be ordered on-line from NOWfoods.com
- Fructose and dextrose can also be used in place of table sugar as a sweetener and for an added source of energy. These work better when added to foods or beverages, but can be used to bake with; recipes will just need to be adjusted.
Patients Diagnosed With GSID

- **Starting Solids**
  - Start as soon as age appropriate; don’t delay
  - If possible, start Sucraird® (sacrosidase) Oral Solution therapy when starting solids
    - Sucraird® 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

Please see Sucraird® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraird® may cause a serious allergic reaction.
Sucraid® (sacrosidase) Oral Solution

Dosing and Administration

- Sucraid® must be kept refrigerated (comes from a specialty pharmacy; delivered to patient’s door)

- 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; heat will diminish the efficacy of the enzyme
    - Do not mix with juice or sucrose containing beverages

- Administer Sucraid® with every meal or snack:
  - Take ½ at beginning of meal/snack and other ½ during meal/snack

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Sucrase-Isomaltase Complex is composed of 2 subunits:

1. **Sucrase** digests:
   - Sucrose → Glucose and Fructose
   - Maltose → Glucose and Glucose (α-1,4)
   - Maltotriose → Glucose, Glucose, and Glucose (α-1,4)

2. **Isomaltase** digests:
   - Isoamaltose → Glucose and Glucose (α-1,6)
   - Maltose → Glucose and Glucose (α-1,4)
   - Maltotriose → Glucose, Glucose, and Glucose (α-1,4)
   - Limit dextrins → Glucose (α-1,4 and α-1,6)

Sucraid® (sacrosidase) Oral Solution is an enzyme replacement therapy for the treatment of 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.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
**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 a 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 can be accessed online at sucraid.net/pi.pdf.
Patients Diagnosed With GSID

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

Please see Sucraïd® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraïd® may cause a serious allergic reaction.
Why Do Sucraid® (sacrosidase) Oral Solution Patients Need to See a Registered Dietitian?

- 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 them.

- 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

- Patients 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.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Adult Diet Guidelines for Patients Diagnosed With GSID

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Choose Foods That Are Low in Sucrose and Starch

<table>
<thead>
<tr>
<th>Dairy</th>
<th>Vegetables</th>
<th>Fruits</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cream cheese</td>
<td>Artichokes</td>
<td>Apples</td>
</tr>
<tr>
<td>Half and half</td>
<td>Asparagus</td>
<td>Blueberries</td>
</tr>
<tr>
<td>Hard cheeses</td>
<td>Broccoli</td>
<td>Strawberries</td>
</tr>
<tr>
<td>Cheddar cheese</td>
<td>Carrots</td>
<td>Grapes</td>
</tr>
<tr>
<td>Swiss cheese</td>
<td>Cabbage</td>
<td>Jujubes</td>
</tr>
<tr>
<td>Feta cheese</td>
<td>Celery</td>
<td>Kiwi</td>
</tr>
<tr>
<td>Cheddar cheese</td>
<td>Corn</td>
<td>Pears</td>
</tr>
<tr>
<td>Sour cream</td>
<td>Cream</td>
<td>Peaches</td>
</tr>
<tr>
<td>Whipping cream</td>
<td>Cucumber</td>
<td>Pineapple</td>
</tr>
</tbody>
</table>

**AVOID Foods High in Starch**

- Baked goods
- Biscuits
- Crackers
- Gluten-Free starches & grains
- Grains
- Pasta

**Notes:** These guidelines are intended as a general guide only. Every individual's situation is different. Dietary advice should be discussed in consultation with a physician and registered dietitian.

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Do not use Sucraid® (sacrosidase). Oral ingestion with patients known to be hypersensitive to yeast, yeast products, prebiotics, or gluten 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, mouth, or throat. Some patients who take Sucraid® may experience worsening of diabetes, symptoms of liver disease, and/or enhanced blood glucose levels. You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch or call 1-800-FDA-1088.

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Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Call SucraidASSIST™ at 1-800-705-1962 for all of your Sucraid® (sacrosidase) Oral Solution needs:

1. Order Sucraid® (each patient assigned a case manager)
2. Consent forms
3. Diagnostic
4. Dietitian*
5. Peer Coach
6. AE/Clinical
7. All other

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Nutrition Topics to Cover

- What is sucrose?
- What is starch?
- What are acceptable alternatives to cow’s milk?
- How much sucrose/starch?
- How do I read a food label?
- What about lactose, fructose?
- What artificial sweeteners are acceptable?
- Is gluten-free ok?
- What about low FODMAPS or SCD?
- How do I meet calorie needs?
- Where do I buy fructose and dextrose?
- Do I need a multivitamin? What kind?
- Etc…
Grams of Sucrose Per Meal / Per Day?

- Compliance with a sucrose-free diet is very difficult.
- After 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 dependent on the individual.

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Grams of Starch Per Meal / Per Day?

- Although Sucrardin® (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.

- Ann McMeans, RD suggests 120 grams/day may be tolerated.

- The type of starch may allow more grams per day (i.e., higher fiber, low GI, adding fat and/or protein with starch, cooking method such as pasta al dente).

- Chewing food well – exposing to amylase longer.

- Some patients take over-the-counter “starch enzymes.”

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Please see Sucrardin® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucrardin® may cause a serious allergic reaction.
# How Much Sucrose and Starch? 6-year-old

<table>
<thead>
<tr>
<th>Nutrient</th>
<th>US AVG (NHANES 2011-2012)</th>
<th>CSID (OBS study 2015)</th>
<th>DRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Energy (kcals)</td>
<td>1983</td>
<td>?</td>
<td></td>
</tr>
<tr>
<td>CHOs (g)</td>
<td>269</td>
<td>?</td>
<td>130</td>
</tr>
<tr>
<td>CHOs (% of kcals)</td>
<td>55</td>
<td>?</td>
<td>45 - 65</td>
</tr>
<tr>
<td>Total Sugars (g)</td>
<td>130</td>
<td>?</td>
<td>&lt;25% added sugars</td>
</tr>
<tr>
<td>Sucrose (g)</td>
<td>40 (8% of kcals)&lt;sup&gt;1&lt;/sup&gt;</td>
<td>?</td>
<td></td>
</tr>
<tr>
<td>Starch (g)</td>
<td>124 (calculated)</td>
<td>?</td>
<td></td>
</tr>
</tbody>
</table>

- **US AVG**: 6 year old, averaged male and female from NHANES 2011-12
- **CSID**: Observational study (N=49), 2015
- **Recommendations**: DRIs

---

Nutrient Data for Sucrose and Starch

- Sucrose and starch data are not a part of most nutrient databases. USDA has limited information, but continues to add these nutrients to their database.
- University of Minnesota Nutrition Data System for Research (NDSR) has complete sucrose and starch data.
- Total carbohydrates and total sugars are on the Nutrition Facts Label.
- Sources of sugar and starch are listed on the ingredient label.
- Detective work.
- One estimate for starch content:

\[
\text{Total CHOs} - \text{total sugars} - \text{fiber} = \text{starch}
\]
Sweeteners

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

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

- **Sweeteners to Avoid:**
  - Beet sugar
  - Brown sugar
  - Cane juice, cane sugar
  - Caramel
  - Coconut sugar
  - Confectioner’s sugar
  - Date sugar
  - Maple syrup
  - Molasses
  - Raw sugar
  - Sucrose
  - Sugar
Starch Ingredients That May Not Be Tolerated

- Limit dextrins
- Corn syrup
- Corn syrup solids
- Maltodextrin
- Modified tapioca starch
- Glucose polymers

(a) Two forms of starch

Amylose
only alpha 1-4
glycosidic bonds

Amylopectin
alpha 1-4 and
alpha 1-6
glycosidic bonds
### Fruit Juice

<table>
<thead>
<tr>
<th>JUICE (4 fluid ounces)</th>
<th>SUCROSE</th>
<th>STARCH</th>
<th>GLYCEMIC INDEX</th>
<th>FRUCTOSE</th>
<th>GLUCOSE</th>
<th>MALTOSE</th>
<th>LACTOSE</th>
</tr>
</thead>
<tbody>
<tr>
<td>JJ APPLE</td>
<td>1.6</td>
<td>0</td>
<td>39</td>
<td>7.1</td>
<td>3.3</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>JJ GRAPE</td>
<td>0.8</td>
<td>0</td>
<td>50</td>
<td>8.2</td>
<td>5.9</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>LIBBY'S PEAR</td>
<td>0.3</td>
<td>0</td>
<td>64</td>
<td>10.7</td>
<td>7.3</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>ORANGE J C, fresh</td>
<td>5.0</td>
<td>0</td>
<td>46</td>
<td>2.8</td>
<td>2.6</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>PINEAPPLE J C</td>
<td>1.9</td>
<td>0</td>
<td>46</td>
<td>4.8</td>
<td>5.8</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>PRUNE J C</td>
<td>0.3</td>
<td>0</td>
<td>29</td>
<td>10.1</td>
<td>7.0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>APPLE-GRAPE DRINK</td>
<td>7.4</td>
<td>0</td>
<td>67</td>
<td>3.3</td>
<td>3.7</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>TOMATO JUICE</td>
<td>0.3</td>
<td>0</td>
<td>33</td>
<td>1.9</td>
<td>1.6</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>
Allowed or Not Allowed On **Elimination Phase** of a **Low-Sucrose, Low-Starch Diet**

- Apple
- Olives
- Banana
- Cracker
- Tomato
- Eggs
- Cottage cheese
- Nuts
- Grapes
- Blueberries
- Butter
- Gluten-Free
- Green beans
- Black-eyed peas
- Yogurt
Think About the Toddler With Chronic Diarrhea

- Started having 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®
## Formula Comparison

<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>EleCare® (Abbott)</td>
<td>amino acids</td>
<td>corn syrup solids</td>
<td>safflower oil, MCT, soy oil</td>
</tr>
<tr>
<td>Neocate Junior® (Nutricia)</td>
<td>amino acids</td>
<td>corn syrup solids</td>
<td>vegetable oil, canola oil, safflower oil</td>
</tr>
<tr>
<td>Splash® (Nutricia)</td>
<td>amino acids</td>
<td>maltodextrin, sugar</td>
<td>coconut oil, canola oil, sunflower oil</td>
</tr>
<tr>
<td>Peptamen Junior® (Nestle)</td>
<td>hydrolyzed whey protein</td>
<td>maltodextrin, sugar, cornstarch</td>
<td>MCT, soybean oil, canola oil</td>
</tr>
</tbody>
</table>
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
  - Normal lactase
  - Normal mucosa
Treatment Plan – Diagnosis of SI Deficiency is Now Confirmed

- Consider changing formula to RCF® (Abbott) or KetoCal® (Nutricia)
- Diet modification and Sucraid® (sacrosidase) Oral Solution therapy

<table>
<thead>
<tr>
<th>FORMULA</th>
<th>PROTEIN SOURCE</th>
<th>CARB SOURCE</th>
<th>FAT SOURCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>RCF® (Abbott)</td>
<td>soy protein isolate</td>
<td>none</td>
<td>safflower oil, soy oil, coconut oil</td>
</tr>
<tr>
<td>% of Kcals</td>
<td>14%</td>
<td>0</td>
<td>86%</td>
</tr>
<tr>
<td>KetoCal 4:1® (Nutricia)</td>
<td>milk protein (casein, whey)</td>
<td>com syrup solids</td>
<td>vegetable oil</td>
</tr>
<tr>
<td>% of Kcals</td>
<td>8.2%</td>
<td>3.1%</td>
<td>88.7%</td>
</tr>
</tbody>
</table>

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Think About the Adult Currently Being Treated For IBS

- You’ve tried medications and various diets

- Patient finds some relief with Low FODMAPs diet
  - But still has lingering symptoms: diarrhea, abdominal pain, gas and bloating
  - And you believe they are compliant with the diet

- Consider that this patient may really have Sucrase-Isomaltase (SI) Deficiency
  - Recommend testing for SI deficiency
  - Reduce sucrose and starch from their current diet
  - Consider a trial of Sucraid® (sacrosidase) Oral Solution therapy and diet modification

Please see Sucraid® Important Safety Information on slides 25-27 and Full Prescribing Information passed out at the beginning of this presentation. Sucraid® may cause a serious allergic reaction.
## Differences Between Elimination Diets

<table>
<thead>
<tr>
<th></th>
<th>Low FODMAPs</th>
<th>Low Sucrose, Low Starch</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Dairy</strong></td>
<td>No (some cheese)</td>
<td>Yes</td>
</tr>
<tr>
<td><strong>Fat</strong></td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td><strong>Protein (meats)</strong></td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td><strong>Starch/grains</strong></td>
<td>Yes (selected)</td>
<td>No</td>
</tr>
<tr>
<td><strong>Fruits</strong></td>
<td>Yes (low fructose)</td>
<td>Yes (low sucrose)</td>
</tr>
<tr>
<td><strong>Vegetables</strong></td>
<td>Yes (selected)</td>
<td>Yes (low starch)</td>
</tr>
<tr>
<td><strong>“Sugars”</strong></td>
<td>No fructose; Yes (sugar, syrup, jelly)</td>
<td>No sucrose; Yes (fructose, dextrose)</td>
</tr>
</tbody>
</table>
THE FUTURE: Sucrase-Isomaltase Deficiency

- Genetic Prevalence Screening study (CSIDgps.com)
  - We believe SI deficiency is more prevalent than previously thought

- Observational study of patients on Sucraid® (sacrosidase) Oral Solution
  - To better understand diet, GI symptoms, Sucraid® usage, and quality of life for those with SI deficiency

- Adult Sucraid® trial
  - Sucraid® vs. placebo

- Publications and educational development

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
In Summary: Consider Genetic SI Deficiency

- When reviewing diet history consider carbohydrate sources (sucrose and starch)
- Ask about foods that trigger symptoms
- **Is Sucrase-Isomaltase Deficiency a possibility?**
  - Share your suspicions with the patient’s doctor and suggest further testing:
    - Disaccharidase assay by endoscopy (especially if already planning to scope)
    - A Sucraid® (sacrosidase) Oral Solution therapeutic trial
    - Diet modifications (sucrose and starch)
    - Additional supportive tests:
      - Sucrose Intolerance Hydrogen Breath Test (SIHBT)
      - SI Genetic Test

Please see Sucraid® (sacrosidase) Oral Solution Important Safety Information and details on accessing the Full Prescribing Information in slides 30-32. Sucraid® may cause a serious allergic reaction.
Questions?

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919-832-0404
800-705-1962 #4

Thank you!