# ANMC Congenital Sucrase-Isomaltase Deficiency (CSID) Diagnosis and Treatment Guidelines

**Updated 8/19/20**

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| - It is estimated that 3-10% of Alaska Native people have CSID, but exact prevalence is unknown  
- There is a mutation present in people from Arctic regions that results in a complete lack of enzyme activity  
- Higher prevalence of CSID is seen along the western/northern coasts of the state, especially Yup’ik and Inupiaq people  
- CSID is an autosomal recessive (homozygous) disorder, but heterozygote carriers may also be symptomatic  
- ANTHC has a CSID Program to provide education and support for patients, families, and providers*  | - Osmotic diarrhea  
- Abdominal pain & distention  
*Symptoms vary between affected individuals depending on the amount of sucrose and/or starch consumed and other unknown factors*  
| - Genetic screen (blood) through Fulgent Genetics  
  - Click [here](#) for test requisition and [here](#) for consent form  
  - In the Test Requested box write “SI gene c.273_274delAG p.Gly92Leufs*8”  
  - Cost: $200  
  - 2-3 week turnaround for results  |
| - Can onset of symptoms be traced back to introduction of table foods? Breast and most formula fed infants are asymptomatic  
- Is there a family history of intolerance or “allergy” to sugar?  
- Are symptoms worse when eating store bought foods versus Alaska Native traditional foods?  | - Malnutrition  
- Failure to thrive  |

### Other Considerations

- Genetic screen (blood) through Fulgent Genetics
  - Click [here](#) for test requisition and [here](#) for consent form
  - In the Test Requested box write “SI gene c.273_274delAG p.Gly92Leufs*8”
  - Cost: $200
  - 2-3 week turnaround for results

### Nutritional Considerations

- Avoid and/or limit sucrose (table sugar) and starch
  - The Alaska Native traditional diet is excellent for managing symptoms of CSID since it is naturally low in sucrose and starch
  - Refer patient to a Registered Dietitian
    - If no RD available, refer to CSID Program Coordinator
  - CSID education materials for patients/families are available. Contact the CSID Program Coordinator for copies.

### Infant Formulas

- Breast milk and standard lactose based formulas, such as Similac® Advance® or Enfamil® Infant, are typically tolerated
- Most other formulas contain sucrose and/or starch and are generally not tolerated

### Medication Management²

- Sacrosidase (Sucraid®): enzyme replacement for sucrose
  - Taken with each meal and snack that contain sucrose
  - Dosage is weight dependent
  - Requires refrigeration
  - Most private insurance covers Sucraid®
  - For Medicaid or IHS recipients, refer patient to ANTHC’s Tribally Sponsored Health Insurance Program (TSHIP). See TSHIP section

### Prescribing Sucraid®

- If prescribing for patients located in the Anchorage Service Unit click [here](#) for instructions
- If prescribing for patients in the Utqiagvik, Kotzebue, and Nome Service Unit check for Sucraid® availability with the local pharmacy

### Tribally Sponsored Health Insurance Program (TSHIP)

- ANTHC partners with THOs to sponsor eligible individuals for health insurance coverage
- Covers the cost of Sucraid®
- 2-6 week period between enrollment and effective coverage
- Does not provide retroactive coverage
- Refer patient to TSHIP by calling 729-2935. A Health Benefits Specialist will work with the patient for eligibility verification and enrollment

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*For questions or assistance, contact ANTHC's CSID Outreach and Education Coordinator at 729-3628*

**REFERENCES:**