Sucraid (sacrosidase) Prior Authorization with Quantity Limit Program Summary

POLICY REVIEW CYCLE

Effective Date 02-01-2025

Date of Origin

FDA LABELED INDICATIONS AND DOSAGE

Agent(s)	FDA Indication(s)	Notes	Ref#
Sucraid® (sacrosidase) Oral solution	Oral replacement therapy for treatment of genetically determined sucrase deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID)		1

See package insert for FDA prescribing information: https://dailymed.nlm.nih.gov/dailymed/index.cfm

CLINICAL RATIONALE

CSID	Congenital sucrase-isomaltase deficiency (CSID) is a rare, chronic, autosomal recessive disorder characterized by the absence or deficiency of the enzymes sucrase and isomaltase.(3) Patients with CSID have two defective copies of the sucrase-isomaltase (SI) gene. The SI enzyme complex is naturally produced in the brush border lining of the small intestine and assists in the breakdown of certain sucrose and products of starch digestion (dextrins). When sucrase-isomaltase is absent or deficient, non-absorbed carbohydrates enter the distal small intestine and colon where they are fermented, leading to the excessive production of short-chain fatty acids and gases such as hydrogen, methane, and hydrogen sulfide. This in turn can lead to abdominal distension, cramping, pain, excessive flatulence, nausea/vomiting, and osmotic diarrhea. If left untreated, significant sucrase-isomaltase deficiency (SID) can result in inadequate growth and failure to thrive in children as well as weight loss in adults.(4) The gold standard for the diagnosis of CSID remains small intestinal biopsy specimens assayed for lactase, sucrase, isomaltase, and maltase activity. Criteria to make the diagnosis of CSID include normal small bowel morphology in the presence of markedly reduced or absent sucrase activity, isomaltase activity varying from zero to full activity, and reduced maltase activity. Lactase activity can be normal or reduced in children with a sucrase:lactase ratio of less than 1.0. Genetic sequencing of the SI gene can identify homozygous and compound heterozygous mutations responsible for CSID. A number of noninvasive diagnostic tests can also help establish the diagnosis, including the sucrose challenge test, lactose breath test, and hydrogen-methane breath test. However, many of these tests have limitations including false-positive results, false-negative results, and lack of validation data.(2) Previously, treatment of CSID has required lifelong adherence to a sucrose-free diet.(2-4) Data suggest that even after diagnosis and dietary treatment, major gastrointestinal symptoms persists, and there is a high frequency of decreased weight for height and age in these patients.(5) Treatment has improved considerably with the availability of enzyme replacement therapy (sacrosidase) which has allowed

	consumption of a more normal diet and decreased the high incidence of chronic gastrointestinal problems.(2-5) Access to a physician or dietician who is knowledgeable about CSID is essential for guiding patients and their families.(4)
Safety	Sucraid is contraindicated in patients known to be hypersensitive to yeast, yeast products, glycerin (glycerol), or papain.(1)

REFERENCES

Number	Reference
1	Sucraid prescribing information. QOL Medical, LLC. December 2023.
2	Treem WR. Clinical Aspects and Treatment of Congenital Sucrase-Isomaltase Deficiency. *Journal of Pediatric Gastroenterology and Nutrition*. 2012;55(S2). doi:10.1097/01.mpg.0000421401.57633.90
3	Congenital Sucrase-Isomaltase Deficiency - Symptoms, Causes, treatment | NORD. National Organization for Rare Disorders. https://rarediseases.org/rare-diseases/disaccharide-intolerance-i/
4	Congenital Sucrase-Isomaltase Deficiency: What, when, and how? – Gastroenterology & Hepatology. https://www.gastroenterologyandhepatology.net/supplements/congenital-sucrase-isomaltase-deficiency-what-when-and-how/
5	Treem WR, McAdams L, Stanford L, Kastoff G, Justinich C, Hyams J. Sacrosidase Therapy for Congenital Sucrase-Isomaltase Deficiency. *Journal of Pediatric Gastroenterology and Nutrition*. 1999;28(2):137-142. doi:10.1097/00005176-199902000-00008

POLICY AGENT SUMMARY PRIOR AUTHORIZATION

Target Brand Agent(s)	Target Generic Agent(s)	Strength	Targeted MSC	Available MSC	Final Age Limit	Preferred Status
Sucraid	sacrosidase soln	8500 UNIT/ML	M ; N ; O ; Y	N		

POLICY AGENT SUMMARY QUANTITY LIMIT

Target Brand Agent Name(s)	Target Generic Agent Name(s)	Strength	QL Amount	Dose Form	Day Supply	Duration	Addtl QL Info	Allowed Exceptions	Targeted NDCs When Exclusions Exist
Sucraid	Sacrosidase Soln 8500 Unit/ML	8500 UNIT/ML	300	mLs	30	DAYS			

CLIENT SUMMARY – PRIOR AUTHORIZATION

Target Brand Agent Name(s)	Target Generic Agent Name(s)	Strength	Client Formulary
Sucraid	sacrosidase soln	8500 UNIT/ML	Commercial ; HIM ; ResultsRx

CLIENT SUMMARY – QUANTITY LIMITS

Target Brand Agent Name(s)	Target Generic Agent Name(s)	Strength	Client Formulary
Sucraid	Sacrosidase Soln 8500 Unit/ML	8500 UNIT/ML	Commercial ; HIM ; ResultsRx

PRIOR AUTHORIZATION CLINICAL CRITERIA FOR APPROVAL

Module	Clinical Criteria for Approval
PA	**Initial Evaluation** **Target Agent(s)** will be approved when ALL of the following are met: 1. The patient has a diagnosis of congenital sucrase-isomaltase deficiency (CSID) confirmed by ONE of the following: A. Genetic testing of the sucrase-isomaltase (SI) gene indicates a pathogenic mutation **OR** B. Endoscopic biopsy of the small bowel indicates normal small bowel morphology in the presence of decreased (or absent) sucrase activity, isomaltase activity varying from decreased to normal activity, and decreased maltase activity **AND** 2. The prescriber is a specialist in the area of the patient's diagnosis (e.g., gastroenterologist, geneticist, endocrinologist), or the prescriber has consulted with a specialist in the area of the patient's diagnosis **AND** 3. The patient does NOT have any FDA labeled contraindications to the requested agent **Length of Approval:** 3 months NOTE: Quantity Limit applies, please refer to Quantity Limit Criteria. **Renewal Evaluation** **Target Agent(s)** will be approved when ALL of the following are met: 1. The patient has been previously approved for the requested agent through the plan's Prior Authorization process [Note: patients not previously approved for the requested agent will require initial evaluation review] **AND** 2. The patient has had clinical benefit with the requested agent **AND** 3. The prescriber is a specialist in the area of the patient's diagnosis (e.g., gastroenterologist, geneticist, endocrinologist), or the prescriber has consulted with a specialist in the area of the patient's diagnosis **AND** 4. The patient does NOT have any FDA labeled contraindications to the requested agent **AND** **Length of Approval:** 12 months NOTE: Quantity Limit applies, please refer to Quantity Limit Criteria.

QUANTITY LIMIT CLINICAL CRITERIA FOR APPROVAL

Module	Clinical Criteria for Approval
Universal QL	**Quantity Limit for the Target Agent(s)** will be approved when ONE of the following is met: 1. The requested quantity (dose) does NOT exceed the program quantity limit **OR** 2. The requested quantity (dose) exceeds the program quantity limit AND ONE of the following: A. BOTH of the following: 1. The requested agent does NOT have a maximum FDA labeled dose for the requested indication **AND** 2. There is support for therapy with a higher dose for the requested indication **OR** B. BOTH of the following: 1. The requested quantity (dose) does NOT exceed the maximum FDA labeled dose for the requested indication **AND**

Module	Clinical Criteria for Approval
	2. There is support for why the requested quantity (dose) cannot be achieved with a lower quantity of a higher strength that does not exceed the program quantity limit **OR** C. BOTH of the following: 1. The requested quantity (dose) exceeds the maximum FDA labeled dose for the requested indication **AND** 2. There is support for therapy with a higher dose for the requested indication **Length of Approval:** up to 12 months