

QA62 – Sucrase and Maltase Deficiency

QUESTION:

A 10 year old female has been diagnosed with lactase/sucrase deficiency. Mom has received charts of sucrose content of foods based on 100 gram portions. (Not very useable for a lay person) I have converted some to household measurements. Is there any reason why a sugar substitute could not be used? Could almond milk or rice milk be used? Are maltodextrins okay? Is there a web site which would have very up-to-date composition of foods including sucrose? The girl can have 0.2 gm of sucrose in a meal. Is the sucrose deficiency “straight forward” as just limiting sucrose in the foods or is there more to watch out for?

ANSWER:

Congenital sucrase-isomaltase deficiency is an autosomal recessive disorder impairing an individual’s ability to metabolize sucrose and maltose. (Sucrase and isomaltase occur together as a complex enzyme.) Some individuals have only reduced levels of isomaltase, and can tolerate larger amounts of maltose. The disorder is typically diagnosed in infancy, but “milder” cases can remain unidentified or misdiagnosed as “chronic diarrhea.” In general, it is treated by limiting intake of sucrose and maltose.

Most sugar substitutes are appropriate though it is important to look at the individual sweetener’s ingredients. Some sweeteners are derived from maltose and isomaltose. The Calorie Control Council website (<http://www.caloriecontrol.org/lowcal.html>) has more information about specific artificial sweeteners. Maltodextrins are derived from cornstarch and are primarily glucose polymers, so are okay. I could not find data for rice or almond milk, however both rice and almonds have small amounts of sucrose. Lactaid-treated cow’s milk might be more appropriate. The USDA website has a pdf file, <http://www.nal.usda.gov/fnic/foodcomp/Data/Other/herr48.pdf>, that provides data about the sugar content of foods, and foods are presented in 100 gram portions as well as household measures.

Because this child can tolerate some amount of sucrose, it is important to reiterate the message that although a specific food may have a small amount of sucrose, it is likely a better choice than a food that has a larger amount of sucrose.

Lactose intolerance in children is not as common as in adults. It is also sometimes a secondary condition, related to intestinal disturbances and gastroenteritis. (This is common in conditions such as untreated celiac sprue. Often, after treatment is started and intestinal villi heal, lactase is produced.) Lactose is added to some sweeteners (including NutraSweet), so if this child cannot tolerate even small amounts of lactose, foods sweetened with NutraSweet may cause symptoms.

References:

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2. Mayes PA. Digestion and absorption. In: Murray RK, Granner, DK, Mayes PA, Rodwell VW, eds. Harper's Biochemistry, 24th edition. Connecticut: Appleton & Lange. 1996.
3. Treem WR. Congenital Sucrase-Isomaltase Deficiency. *Journal of Pediatric Gastroenterology and Nutrition*, 1995; 21:1-14.