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Introduction to the 8th Starch Digestion Consortium Workshop

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Congenital sucrase-isomaltase deficiency (CSID) is a rare autosomal intestinal disease caused by mutations of the sucrase-isomaltase gene. Patients with CSID have different phenotypes and enzymatic activities associated with sucrase-isomaltase ranging from mild reduction of sucrase activity to complete absence. Low sucrase activity leads to maldigestion of sucrose, resulting in dyspepsia-like symptoms such as chronic diarrhea, abdominal pain, and bloating. The severity of the symptom is related to the amount of sucrase activity and quantity of sucrose ingested. Reduced maltase activity is expected to occur in patients with CSID because both subunits of the SI complex contribute to the total mucosal maltose activity. Indeed, low maltase activity can also lead to maldigestion of starches contributing to the symptoms of dyspepsia. When children are assessed for this gene mutation, duodenal mucosal histology is invariably normal.

Because there are no noninvasive methods for specific confirmation of sucrase-isomaltase deficiency, a novel noninvasive ^{13}C -sucrose labeled substrate has been developed and validated as a sucrase activity breath test for screening and confirmation of CSID. It has been shown that primary sucrase deficiency can be confirmed using this new ^{13}C breath test, as well as the effectiveness of sucrase replacement therapy.

On December 21 and 22, 2011, the 8th Workshop of the Starch Digestion Consortium (SDC) was held at the Children's Nutrition Research Center at the Baylor College of Medicine and Texas Children's Hospital. The theme of the workshop was "50 Years of Progress Since Congenital Sucrase-Isomaltase Deficiency (CSID) Recognition." This was a multidisciplinary workshop that blended clinical medicine with nutritional and food science. The purpose of this workshop was to review progress toward clinical diagnosis and management of sucrase-isomaltase enzyme deficiency during the past 50 years. The nutritional and food technological objectives of the conference were 2-fold: first, to define the role of sucrase-isomaltase enzyme complex in human starch digestion to glucose (α -glucogenesis), and second, by studying sucrase-isomaltase-deficient patients with CSID, envision approaches to botanical and technological alterations in food that will benefit all populations.

More than 50 authors and attendees participated in this workshop from 12 different countries. Eighteen original communications were presented. Special thanks go to QOL Medical, the supplier of Sucraid oral enzyme supplement, for sponsoring this event. This supplement to the *Journal of Pediatric Gastroenterology and Nutrition* reports the

proceedings of this workshop. E. Roseland Klein served as the technical editor of the workshop publication. Beth Mays was responsible for all travel arrangements and the workshop organization. Adam Gillum was responsible for graphic and audio arrangements.

The group of participants standing among the lobby statuary at the SDC/CSID workshop held at the Children's Nutrition Research Center, December 21–22, 2011 is shown in Figure 1. Statues are identified by italics; *Borden* is by Joseph Paderewski (1914–2000) and the others are copies of the work of Elizabet Ney (1833–1907).

FIGURE 1.

From left to right: back row, William Bryant, *Justus Leibig*, Mohammad Chegeni, Peter Zimmer, David Rose, Mark Gilger, Antone Opekun, *Gale Borden*, Christina Lecea; third row, Bruno Chumpitazi, Ronald Scott, William Treem, *Sursum*, Derick Cooper, Zihua Ao, Byung-Hoo Lee; second row, Amy Lin, Beth Mays, William Dupere, Susan Baker, Buford Nichols, Teresa Davis, Bruce Hamaker, Roberto Quezada-Calvillo, Steve Avery; front row, Kyra Jones, Marcia Chaudet, Hassan Naim, Mary Money-Waldman, Veda Nichols, Fatimata Cisse, Meric Simsek, Like Yan, Ann McMeans, Mary Slawson.