Evaluation of Recommendations for Nutrition Management of Classical Galactosemia

SANDY VAN CALCAR PHD, RD ASST. PROFESSOR OF PEDIATRICS SENIOR METABOLIC DIETITIAN BIOCHEMICAL GENETICS PROGRAM, WAISMAN CENTER UNIVERSITY OF WISCONSIN-MADISON

Aim of Project

- Improve consensus for diet treatment of classical galactosemia.
- Why?
 - o All agree galactose restriction is life-saving in infancy
 - o Question: how strict does diet need to be the long-term?
 - o Little agreement between clinic's policies
 - o Need for guidelines, based on CURRENT KNOWLEDGE

Two major projects

- 1. Galactose analysis of two major questionable foods/ingredients:
 - o Sodium and calcium caseinate
 - Aged cheeses
- Samples sent from different locations
- Analyzed 22 products

Two major projects

- 2. Develop a task force of professionals and parent representative to:
- Evaluate current knowledge about diet
- o Identify areas that need further investigation
- Determine educational needs for parents, patients and professionals
- $\circ\,$ Based on what we know now, establish recommendations for diet management
- o Present findings in peer-review journal and for consumers

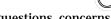
Task Force Members

- Sandy van Calcar, PI
- Physician
 - o Gerry Berry, MD, Children's Hospital of Boston
- Metabolic Dietitians
 - o Laurie Bernstein, Children's Hospital of Colorado
- o Fran Rohr, Children's Hospital of Boston
- Food Scientist
 - o Chris Scaman, University of British Columbia

Task Force members, cont.

- Industry Representative
 - o Steven Yannicelli, Nutricia North America
- Parent Representative
 - o Denise Wilburn
- Prior to meeting: collect research articles, current food lists and educational materials

Topics discussed



- Parent's questions, concerns
- Infant feeding
- Link between diet and long-term outcome
 - o Patient data: dietary challenges and effect on gal-1-P levels
- Food science of galactose
- Results of caseinate and cheese analysis
- Results from patient chart reviews
- Current diet recommendations, materials
- Identify:
- o Research needs
- o Educational needs

Project Products

- Posters at two scientific meetings
- Review article in peer-reviewed professional journal
 - o Review topics from meeting
 - o Food analysis results
 - o Task Force recommendations
 - o Identify future research and educational needs
- Translate recommendations into consumer-friendly educational projects