

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

Two major projects

1. Galactose analysis of two major questionable foods/ingredients:

- 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
- Identify areas that need further investigation
- Determine educational needs for parents, patients and professionals
- Based on what we know now, establish recommendations for diet management
- Present findings in peer-review journal and for consumers

Task Force Members

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

Task Force members, cont.

- Industry Representative
 - Steven Yannicelli, Nutricia North America
- Parent Representative
 - 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
 - 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:
 - Research needs
 - Educational needs

Project Products

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