

## DIET FOR GALACTOSEMIA: LIFE IN THE “GREY ZONE”

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## Big Ticket Items

- We all agree on this....
- Eliminate Dairy Products
  - Milk, buttermilk, cream
  - Yogurt
  - Cheese (most)
  - Whey, nonfat dry milk, milk solids
- Eliminate Organ meats
  - “Meat By-products”

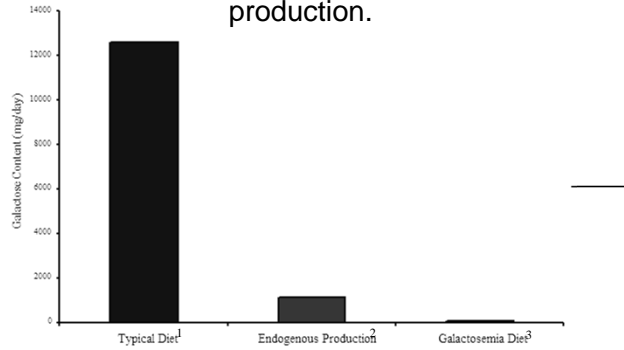
## Recommendations are less clear for...

- Plant Products
  - Fruits and Vegetables
  - Legumes
  - Soy Products
  - Fermented and pickled foods
- Dairy Products
  - Caseinates
  - Aged Cheese

## A Historical perspective....

- Easy diet, good outcome
- Waggoner study (1990) changed this perception
  - Growth delays
  - Ovarian dysfunction
  - Speech abnormalities
  - Learning disabilities, neurological complications
- Studies measuring free galactose in plant foods in 1990's
- Studies measuring endogenous galactose production >> dietary galactose intake (1995 +)
- Studies suggesting that a more liberal galactose intake may not be a problem AFTER infancy (2000+)

### Comparison of the galactose content of a regular diet, galactosemia diet and endogenous galactose production.



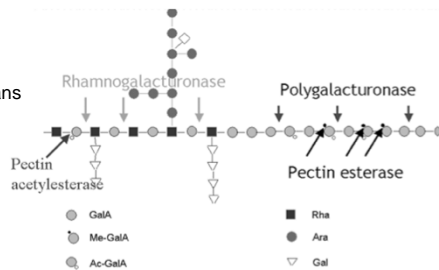
<sup>1</sup> Approximate galactose content of a typical diet including 2 cups of milk and 3 servings of fruits and vegetables with a galactose content >20 mg galactose/100 g food.  
<sup>2</sup> Approximate amount of endogenous galactose produced by an adult with galactosemia.  
<sup>3</sup> Approximate galactose content of a typical galactose-restricted diet including 3 servings of fruits and vegetables with a galactose content >20 mg galactose/100 g food.

### Studies suggesting that a more liberal intake of galactose is OK after infancy

### Galactose in Plants: Free vs Bound

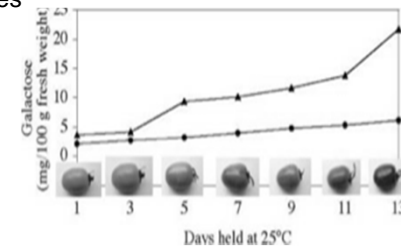
Free Galactose

Bound Galactose in cell walls  
 Alpha 1,4 linkages  
 not digested in humans



### What breaks down bound galactose to release free galactose?

- Ripening
- Longer Storage
- Higher temperatures
- Enzyme treatment
- Heating
- Canning
- Processing



## Fruits and Vegetables

### Studies measure free galactose:

- Tomato sauce = 77 mg in 1/2 cup
- Tomatoes, raw = 23 mg in 1 medium
- Blueberries = 26 mg in 2/3 cup
- Watermelon = 15 mg in 2/3 cup

(Gropper et al., J Am Diet Assn, 2000)

But, what do these numbers mean compared to dairy products?



1 ounce of Milk = 100 mg galactose

Galactosemia, dietary management in the Netherlands, Spa, Belgium, October 2009



5 ½ lbs of Tomatoes = 100 mg galactose

Galactosemia, dietary management in the Netherlands, Spa, Belgium, October 2009



16 ½ lbs. of Apples = 100 mg galactose

Galactosemia, dietary management in the Netherlands, Spa, Belgium, October 2009

## Task Force recommendations

- Fruits and Vegetables YES!!!! OK!!!!
- Besides galactose content, what did we consider?
  - Nutritional value of fruits and vegetables
  - Some clinics eliminate until child is older
    - Early exposure to foods is important



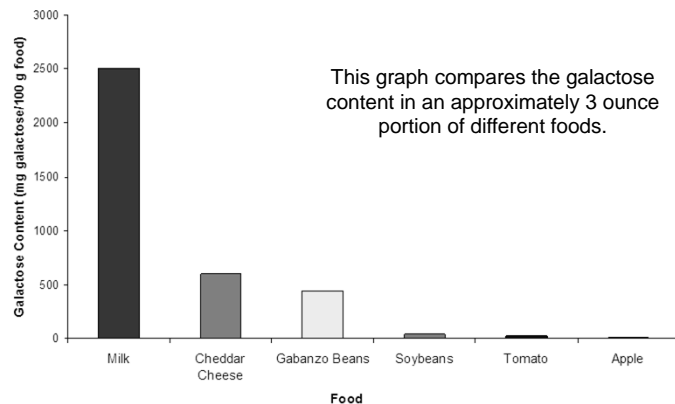
## Legumes (mg free galactose/100 g) Boiled for 2.5 hours, analyzed legumes and water

Garbanzo Beans	444 mg in 2/3 c. cooked
Lima Beans	175 mg
Kidney Beans	150 mg
Lentils	120 mg
Pinto	42 mg

(Acosta & Gross, Eur J. Ped, 1995)



## Comparison of Approximate Amounts of Galactose in Various Foods

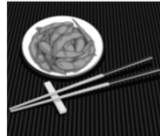


## Some considerations about legumes

- Legumes contain a lot of bound galactose
  - Raffinose, stachyose
- Consider processing used in studies. Other studies analyzing galactose content have found lower values
- Clinics that allow legumes (including garbanzo beans) have not seen an increase in gal-1-P levels
- Task Force Recommendations
  - Allow all legumes
  - Need further analysis, especially for garbanzo beans

## Soy products

- Whole soybeans: 23 mg/100 g cooked
- Soy milk (whole beans): 2.9 - 6.7 mg/100 g
- Formula with soy protein isolate < 2 mg/100g
- Other products
  - \*Soy milk: tofu, soy yogurt, soy cheese
  - \*Soy oil, \*soy lecithin
  - Whole beans: soy flour, flakes, nuts, TSP
  - Soy protein concentrate: Meat analogs
  - Fermented: soy sauce, miso, natto, tempeh



## Soy sauce

- Fermented soy sauce (Kikkomen®)
  - Made by fermenting whole soybeans
  - 170 mg/100 g = 31 mg/Tbsp
  - Tamari, Shoyu, Teriyaki Sauce
- Unfermented soy sauce (LaChoy®)
  - Made from hydrolyzed soy protein
  - Likely less galactose, but not measured

## Task Force recommendations

Soy milk OK.

“Whole soybeans” listed on label

Products made from soy milk OK.

Fat-based soy oil and lecithin OK.

Whole bean products OK.

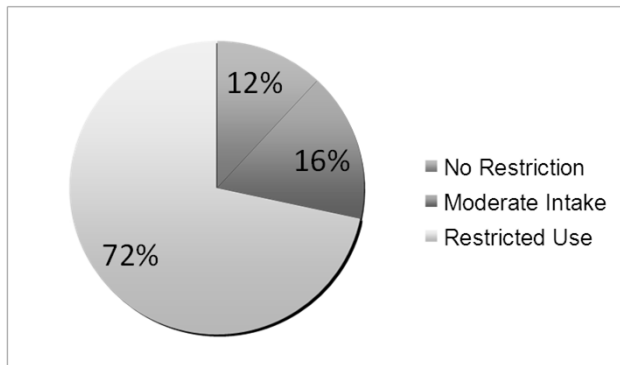
Soy sauce OK.

Need to analyze some of the fermented foods.

## Caseinates

- Sodium and calcium salts of casein
  - Bind water, emulsifier, thickener, “bodying agent”
  - Concern about contamination from lactose
  - Caseinates – confusion with casein?
- Caseinates are “purer” now than in the past
  - “Galactose is not detected”
    - G. Broderick, Professor, Wisconsin Dairy Research Center
- Do we need to continue to avoid caseinate?

### Clinic policies on the restriction of caseinates



### Analysis of Caseinates

	Source	Galactose (mg/100 g)
<b>Sodium Caseinate</b>	US - Wisconsin	95.5
	New Zealand	62.0
	Netherlands *Soy oil, *soy lecithin <b>AVERAGE</b>	< 5 <b>54.2 mg/100 g</b>
<b>Calcium Caseinate</b>	US - Wisconsin	< 5
	New Zealand	40.3
	Netherlands <b>AVERAGE</b>	< 5 <b>16.8 mg/100 g</b>



### What foods contain caseinates?

- Baked products
- Nondairy creamer and products made from this
- “whip cream”
- Non-dairy cheeses (i.e. soy cheese)
- “Muscle-building” protein products
- NOTE:
  - Some of these foods will also have other dairy products
  - Casein is not the same as caseinates
  - Where are caseinates listed on the ingredient list?
- **Task Force Recommendation:** OK to use foods with caseinates, if all other ingredients are acceptable.
  - Casein remains unacceptable

## Galactose in Aged Cheese

- Definitions:

- Mild: Aged 2 - 3 months
- Medium: Aged 4 - 6 months
- Sharp: Aged > 9 months



- Types of cheese considered OK for galactosemia

- Emmentaler, Gruyere, Tilsiter
  - Analyzed brands with Schaerdinger Dairy Association seal (Switzerland)

## Galactose in Aged Cheese

- European study finds undetectable galactose in:

- Jarslberg (Norwegian Emmentaler)
- Italian Parmesan (Parmigiano Reggiano, Grana Padon)
- Hard, aged cheddar (> 1 year)

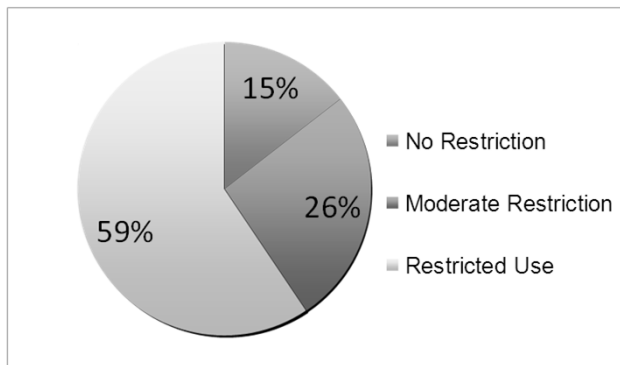
Portnoi and MacDonald, J Hum Nutr Diet, 2009

- Low galactose content ( ave. 3 mg/100 g) in Baby Minibel (Laughing Cow®) Edam and Emmentaler

Portnoi and MacDonald, J Hum Nutr Diet, 2011

- Other studies suggest low levels in Swiss and Romano Cheeses

## Clinic policies on the restriction of Aged Cheese



## Factors that can affect the final galactose content of cheese

- **Bacterial species used for cheese culture**
- **Aging** – galactose generally decreases with age
- **Temperature during aging** - galactose decreases slower at lower temps
- **Salting** – can inhibit the bacteria, and therefore less galactose metabolized
- **Curd washing** – removal of lactose

## Cheese Analysis

- Question 1: Various cheeses have been analyzed in Europe and found to have undetectable galactose. Do these same types of cheese produced in the US also contain undetectable galactose?

## Cheese Analysis

Type of Cheese	Source	Galactose (mg/100 g)
Gruyere	Switzerland	< 5.1 (below quantitation level)
	US - Wisconsin	< 5.1
Emmentaler	Switzerland	< 5.1
	US - Wisconsin	< 5.1
Tilsiter (not analyzed)	Switzerland	
Swiss	US - Wisconsin	7.4

## Cheese Analysis

- Question 1: Various cheeses have been analyzed in Europe and found to have undetectable galactose. Do these same types of cheese produced in North America also contain undetectable galactose?
- Question 2: Sharp cheddar cheese aged > 1 year has been analyzed in Europe and found to have undetectable galactose. Are sharp cheese's available in North America ok for the galactosemia diet?

## Cheese Analysis – Sharp Cheddar

Product	Manufacturer	Place of Purchase	Galactose (mg/100 g)
Sharp - 1 yr age	Brennan's - WI	Madison	6.6
Seriously Sharp	Cabot - Vermont	Boston	<5.1
Naturally Sharp	Cracker Barrel – Kraft IL	Boston	60.4
Extra Sharp White	Cracker Barrel – Kraft IL	Madison	16.5
Sharp	Tillamook - Oregon	Portland, OR	14.7
Sharp	Kraft - Illinois	Denver	104.3
Sharp	JS - Wisconsin	Madison	7.4
Fromage Cheddar	Kraft - Ontario	Vancouver BC	40.3
Fromage Cheddar Fort	Lucerne Foods - Calgary	Vancouver BC	< 5.1

**AVERAGE: Kraft = 50 mg Local = 8 mg**



## Cheese Analysis

- Question 1: Various cheeses have been analyzed in Europe and found to have undetectable galactose. Do these same types of cheese produced in North America also contain undetectable galactose?
- Question 2: Sharp cheddar cheese aged > 1 year has been analyzed in Europe and found to have undetectable galactose. Are sharp cheese's available in North America ok for the galactosemia diet?
- Question 3: What about parmesan cheese?
- Question 4: What about medium and mild cheese?

## Cheese Analysis: Parmesan

Form	Manufacturer	Galactose (mg/100 g)
Powder	Kraft - Illinois	26.3
Block – 10 mo aged	Brennan's - Wisconsin	< 5.1

## Cheese Analysis: Medium and Mild Cheddar

Type	Manufacturer	Galactose (mg/100 g)
Medium – 6 mo aged	Brennan's - Wisconsin	< 5.1
Mild – 3 mo aged	Brennan's - Wisconsin	5.6
Mild – Yogurt base	Cabot - Vermont	< 5.1

## Cheese Analysis

- Question 1: Various cheeses have been analyzed in Europe and found to have undetectable galactose. Do these same types of cheese produced in North America also contain undetectable galactose?
- Question 2: Sharp cheddar cheese aged > 1 year has been analyzed in Europe and found to have undetectable galactose. Are sharp cheese's available in North America ok for the galactosemia diet?
- Question 3: What about parmesan cheese?
- Question 4: What about medium and mild cheese?
- Question 5: Does "lactose-free" mean "galactose-free"?

## Cheese Analysis: "Lactose-free"

Type	Manufacturer	Galactose (mg/100 g)
Naturally Sharp Cheddar	Cracker Barrel – Kraft IL	60.4
Extra Sharp White Cheddar	Cracker Barrel – Kraft IL	16.5
Sharp Cheddar	Kraft - Illinois	104.3
Mild Cheddar - yogurt based	Cabot – Vermont	< 5.1
Sharp Cheddar	Cabot - Vermont	< 5.1

Of all cheese analysis, only one contained detectable lactose

## Conclusions about Aged Cheese

- European and US brands of Emmentaler and Gruyere are OK
- Parmesan cheese –
  - Italian brands OK
  - US brands of brick parmesan aged > 10 months OK
  - Powder contains some galactose, but consider amount used.
- Sharp cheddar cheese – variable results. Local may be lower than large national brands
- Mild and medium cheddar cheese – may not be that much greater in galactose than sharp cheddar
- “Lactose-free” doesn’t necessarily mean “galactose-free”

## Foods and Ingredients that Contain Galactose

-Understanding Galactosemia: A Diet Guide, 2010

- **Butter**
- **Buttermilk**
- **Buttermilk Solids**
- **Casein**
- **Caseinates**
- **Cheese** - including cottage cheese, cream cheese and other cheese-based products **EXCEPT Gruyere, Emmentaler, brick Parmesan**
- **Cream**
- **Curds**
- **Dry milk**
- **Dry Milk Protein**
- **Dry Beans and Peas**
- **Garbanzo Beans** - can also be called chickpeas
- **Ghee** - a clarified butter commonly used in Indian cooking
- **Hydrolyzed whey protein** - when made from casein or whey
- **Ice cream**
- **Lactalbumin** - can also be called milk albuminate
- **Lactose**
- **Lactoglobulin**
- **Margarine** - a few diet margarines or kosher margarines do not contain milk products and are acceptable.
- **Milk**
- **Milk chocolate**
- **Milk solids**
- **Nonfat dry milk**
- **Nonfat dry milk solids**
- **Nonfat milk**
- **Organ meats** - these include liver, heart, kidney, brains, sweetbreads, and pancreas. These are often listed as “**meat byproducts**” on labels.
- **Sherbet** - contains nonfat dry milk. This is different from *sorbet* which is more like a fruit ice and is often acceptable.
- **Sour cream**
- **Whey and whey solids**
- **Yogurt**

## Some things to think about....

- Diet is life-saving for infants, but we need to realize that diet alone does not change long-term outcome.
- Are we affecting quality of life with excessive restrictions that may not be necessary?
- Berry & Elsas, J Inherit Metab Dis, 2011  
“Some physicians are concerned that we have transformed galactosemia into a progressive disease through the very use of a chronic strict diet therapy that limits galactose intake and thus creates further deficiency of UDP-galactose and UDP-glucose in some target tissues”

## Diet Monitoring

- Mutation studies
  - Homozygous Q188R considered most “classical”
  - Help predict dietary recommendations?
- RBC Galactose-1-Phosphate
  - Not very sensitive to diet changes
  - Goal: Less than 3.5 mg/dl (or 4.0 mg/dl)
  - Little relationship between Gal-1-P and long-term outcome
- Most will have “baseline” range of Gal-1-P
  - For monitoring diet: Must see major changes in Gal-1-P, small changes do not reflect diet
- Urine or plasma galactitol

## Conclusions

- We need to continue to reevaluate our diet recommendations for classical galactosemia.
- We need standardized diet recommendations – they may change as more research becomes available, but mixed messages are not helping families.
- **Thank you!**
  - Galactosemia Foundation support and funding
  - Galactosemia Task Force Members