



New International Diet Guidelines for Galactosaemia





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GUIDELINES

International clinical guideline for the management of classical galactosemia: diagnosis, treatment, and follow-up

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On behalf of the Galactosemia Network (GalNet)

Poll 1: Since the 2017 guidelines have you made any changes to your diet?

1)No

2)Yes

Infant Formula

Recommendation #4 (++)

Clinicians should immediately commence a galactose-restricted diet (e.g., soy-based, casein hydrolysate or elemental formula) if classical galactosemia is suspected in an infant, without waiting for confirmation of the diagnosis.



Poll 2: Infants and children; How do you manage your diet?



- 1) Avoid milk and some vegetables and legumes
- 2) Avoid milk products, and strictly check labels and foods in restaurants
- 3) Avoid milk products, but not strict with checking labels and foods in restaurants
- 4) Avoid milk, but occasionally may have a baked product with milk in (for example bread)
- 5) Don't restrict anything
- 6) Not applicable

Poll 3: Adults; How strict are you with the diet?



- 1) Avoid milk and some vegetables and legumes
- 2) Avoid milk products, and strictly check labels and foods in restaurants
- 3) Avoid milk products, but not strict with checking labels and foods in restaurants
- 4) Avoid milk, but occasionally may have a baked product with milk in (for example bread)
- 5) Don't restrict anything
- 6) Not applicable

Diet strictness

Recommendation #5 (expert opinion, +)

We recommend treating patients with CG with a life-long galactose-restricted diet that only eliminates sources of lactose and galactose from dairy products, but permits galactose from non-milk sources that contribute minimal dietary galactose. Within this definition we accept that small amounts of galactose are present in specific mature cheeses and caseinates. At present there is insufficient evidence to support a specific age-related recommendation for the quantity of galactose allowed in the diet.



Diet strictness

Rubio-Gozalbo et al. *Orphanet Journal of Rare Diseases*
<https://doi.org/10.1186/s13023-019-1047-z>

(2019) 14:86

Orphanet Journal of
Rare Diseases

RESEARCH

Open Access

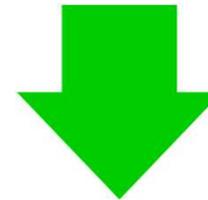
The natural history of classic galactosemia: lessons from the GalNet registry



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A restriction
of milk
sources only

A restriction of
milk sources +
vegetables and
fruit



Better
neurological
outcomes



Poll 4: Which food has the highest galactose content?



1) 100mls of cows milk



2) 100g of cows butter



3) 100g of butter oil / milk fat



4) 100g serving of vegetables

Poll 4: Which food has the highest galactose content?



1) 100mls of cows milk = 2400mg of galactose



2) 100g of cows butter = 685mg of galactose

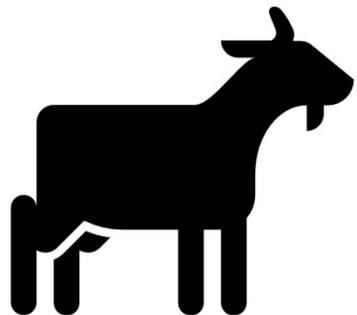
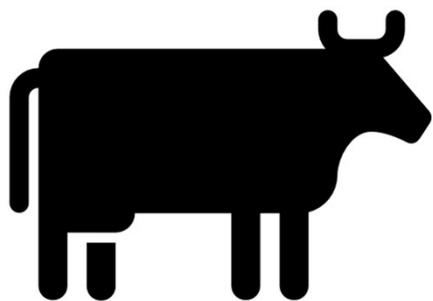
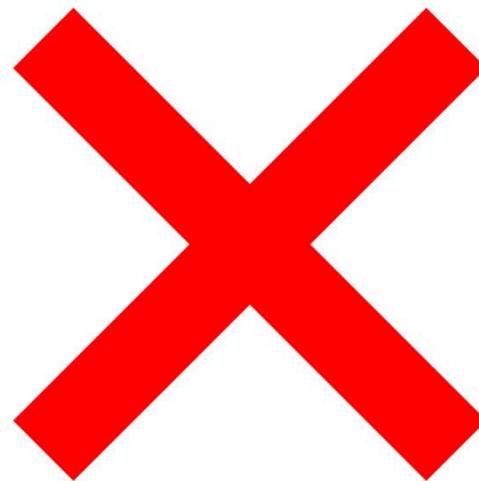


3) 100g of butter oil / milk fat = <math><0.05-2.3\text{ mg}</math> of galactose



4) 100g serving of vegetables = ~9mg of galactose

Milk and Milk products



14 allergens

If your product contains any of the main 14 allergens as an ingredient or processing aid, it must be included on the label.

The 14 main allergens are:

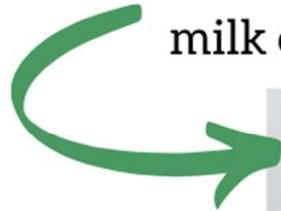
- celery
- cereals containing gluten – including wheat (such as spelt and Khorasan), rye, barley and oats
- crustaceans – such as prawns, crabs and lobsters
- eggs
- fish
- lupin
- milk
- molluscs – such as mussels and oysters
- mustard
- tree nuts – including almonds, hazelnuts, walnuts, brazil nuts, cashews, pecans, pistachios and macadamia nuts
- peanuts
- sesame seeds
- soybeans
- sulphur dioxide and sulphites (if they are at a concentration of more than ten parts per million)

EU Allergen Labelling Laws



Checking the list of ingredients for milk

The ingredients list does not have milk on it so is **safe** to eat



Our ingredients

Wheat Flour, Olive Oil (7%), **Barley** Malt Extract, Iodised Salt, Yeast, Flavourings.



Allergy advice: For allergens, including cereals containing gluten, see ingredients in **bold**. Also, not suitable for customers with an allergy to sesame, milk or soya due to manufacturing methods.



This ingredients list does have milk on it so it is **not safe** to eat



Our ingredients: **Oats** (43%), Golden Syrup, Chicory Fibre, Yogurt Coating (10%) (Sugar, Palm Kernel Fat, Lactose (Cows' **Milk**), Yogurt Powder (Cows' **Milk**), Emulsifier: **Soya** Lecithin)), Rapeseed Oil, Sultanas (4%), Concentrated Grape Juice, Rice Starch, Sunflower Seeds (2%), Pumpkin Seeds (2%), Chia Seeds (1.5%), Vegetable Glycerine, Blueberries (0.5%), Yogurt Powder (Skimmed Cows' **Milk** Solids, Lactic Starter Culture), Flavourings, Thickener: Xanthan Gum.



Allergy advice: For allergens, including cereals containing gluten, see ingredients in **bold**. Also, not suitable for customers with an allergy to nuts due to manufacturing methods.



Butter oil / Milk Fat



Information

Ingredients

Flour (39%) (**Wheat** Flour, Calcium, Iron, Niacin, Thiamin), Plain Chocolate (30%) [Sugar, Cocoa Mass, Vegetable Fats (Palm, Sal and/or Shea), Butter Oil (Milk), Cocoa Butter, Emulsifiers (**Soya** Lecithin, E476), Natural Flavouring], Vegetable Oil (Palm), Wholemeal **Wheat** Flour (9%), Sugar, Glucose-Fructose Syrup, Raising Agents (Sodium Bicarbonate, Malic Acid, Ammonium Bicarbonate), Salt

Allergy Information

Contains: Milk, Soya, Wheat

Butter oil / Milk Fat



Ingredients

Flour (39%) (**Wheat** Flour, Calcium, Iron, Niacin, Thiamin), **Milk** Chocolate (30%) [Sugar, Cocoa Butter, Cocoa Mass, Dried Skimmed **Milk**, Dried Whey (**Milk**), Butter Oil (**Milk**), Vegetable Fats (Sal and/or Shea, Palm), Emulsifiers (**Soya** Lecithin, E476), Natural Flavouring], Vegetable Oil (Palm), Wholemeal **Wheat** Flour (9%), Sugar, Glucose-Fructose Syrup, Raising Agents (Sodium Bicarbonate, Malic Acid, Ammonium Bicarbonate), Salt

Allergy Information

Contains: Milk, Soya, Wheat



Poll 5: Are lactose free products ok to have?

- 1) No
- 2) Yes



Lactose free products

(aimed at people who are lactose-intolerant)

They still
contain
galactose!



Poll 6: Do you allow foods that say on the label 'may contain a source of milk' or 'made in the same factory as milk'

1) No

2) Yes

Allergy Advice

This product is produced in a bakery which uses Milk, Sesame Seeds and Barley Not suitable for someone with Sesame allergy

Allergy Advice

Made in a factory that also handles: Milk, Wheat, Gluten, Barley, Soya, Celery, Mustard

Cross Contamination

There will be very little or no lactose in these products so they will be suitable in the diet.

Allergy Advice

This product is produced in a bakery which uses Milk, Sesame Seeds and Barley Not suitable for someone with Sesame allergy

Allergy Advice

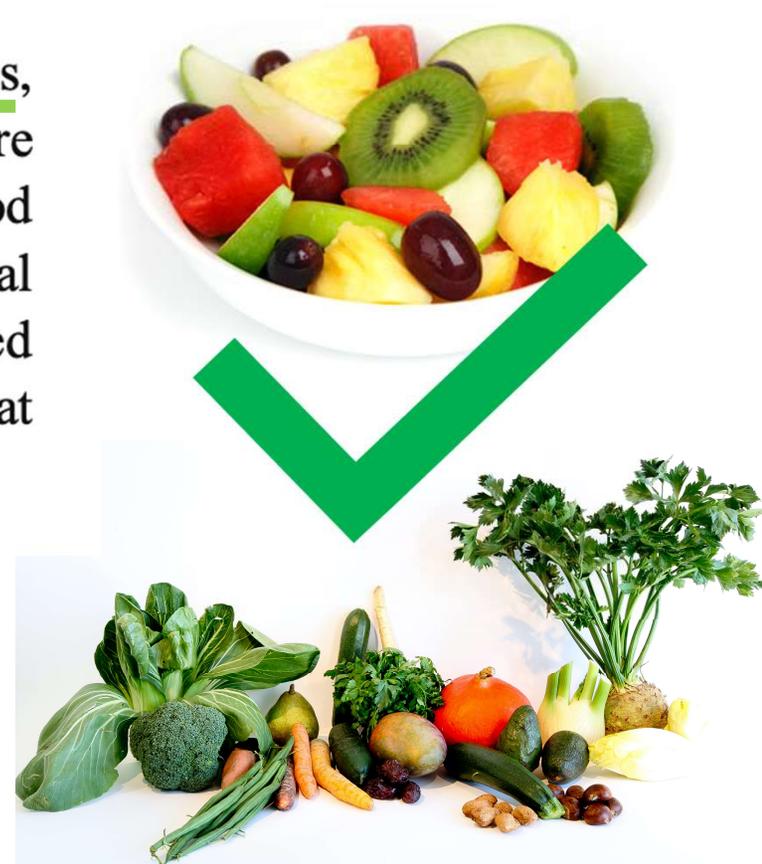
Made in a factory that also handles: Milk, Wheat, Gluten, Barley, Soya, Celery, Mustard



Diet strictness

Recommendation #6 (+)

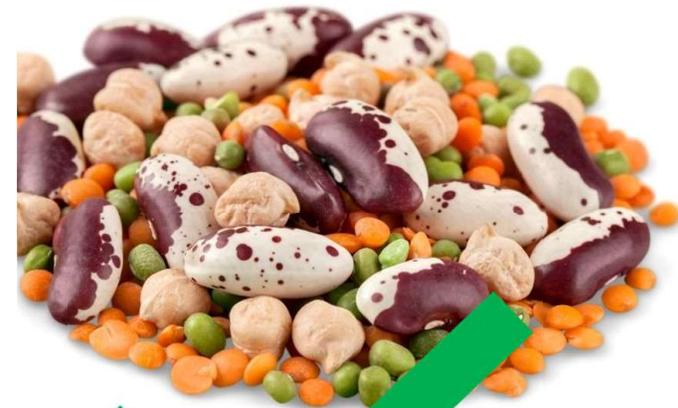
We recommend allowing any amount and type of fruits, vegetables, legumes, unfermented soy-based products, mature cheeses (with galactose content <25 mg/100 g), and the food additives sodium or calcium caseinate, in the diet for classical galactosemia. Although higher in galactose, all fermented soy-based products can be allowed in the small quantities that are typically used in the diet.



Diet strictness

Recommendation #6 (+)

We recommend allowing any amount and type of fruits, vegetables, legumes, unfermented soy-based products, mature cheeses (with galactose content <25 mg/100 g), and the food additives sodium or calcium caseinate, in the diet for classical galactosemia. Although higher in galactose, all fermented soy-based products can be allowed in the small quantities that are typically used in the diet.

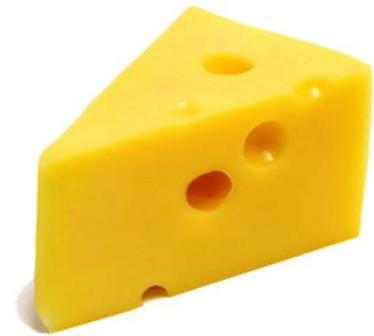


Tofu

Diet strictness

Recommendation #6 (+)

We recommend allowing any amount and type of fruits, vegetables, legumes, unfermented soy-based products, mature cheeses (with galactose content <25 mg/100 g), and the food additives sodium or calcium caseinate, in the diet for classical galactosemia. Although higher in galactose, all fermented soy-based products can be allowed in the small quantities that are typically used in the diet.



Poll 7: Do you allow any cheese in the diet?

- 1) No
- 2) Yes, allow all cheese
- 3) Yes, only certain types of cheese
- 4) Vegan cheese



Cheese

Mature
cheese with a
galactose
content of
less than
25mg / 100g



Cheese with
a galactose
content of
more than
25mg / 100g



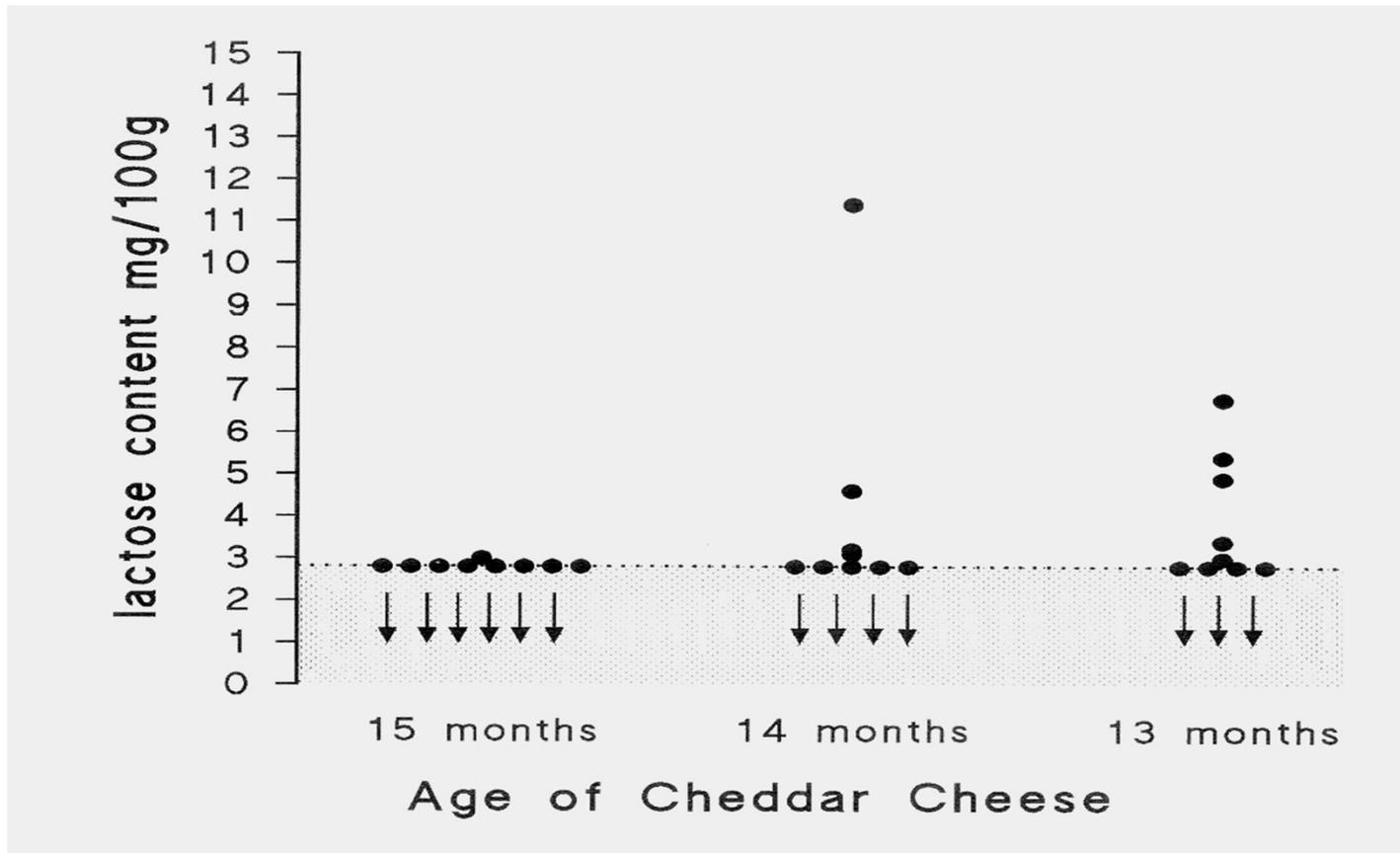
How is lactose lost in cheese

Bacteria in the Starter Culture (added in cheese making) uses up the lactose

As the cheese ages and dries, it loses whey protein and lactose, as it becomes drier



Lactose content of cheddar cheese over time



Poll 8: Do you allow any offal (i.e. kidney, liver, liver pate) in the diet?

1) No

2) Yes

3) In small amounts

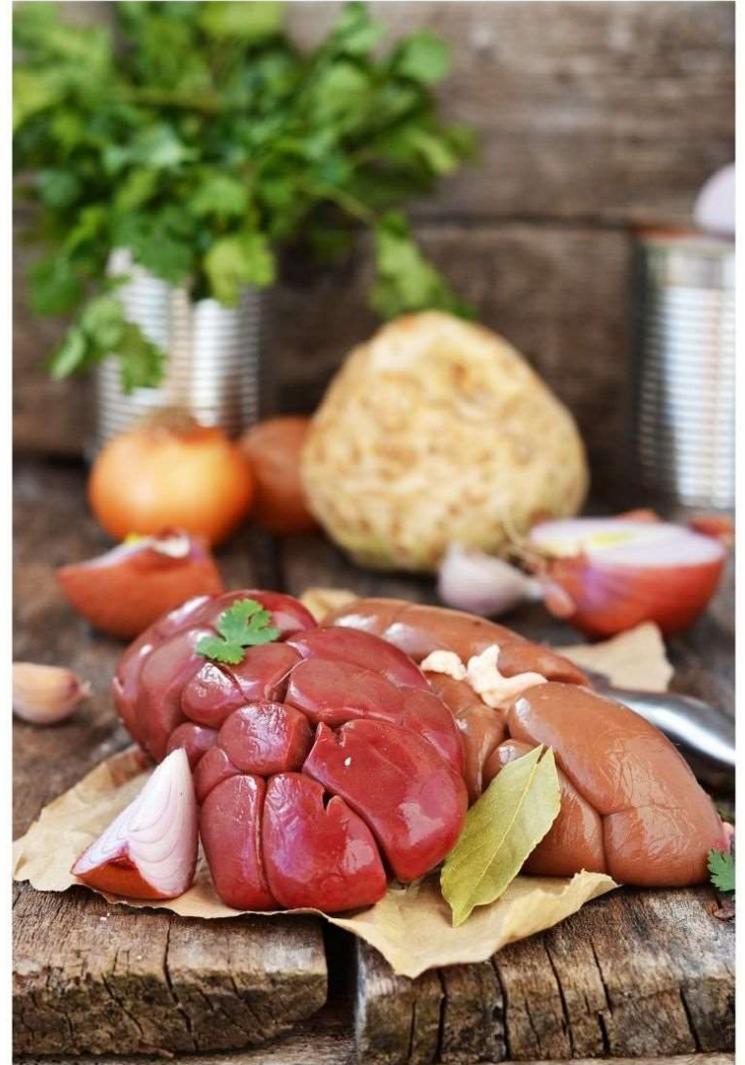


Table 3 Current diet restriction for classical galactosemia (adapted from Bernstein et al, Children's Hospital Colorado in collaboration with the Galactosemia Foundation Task Force) (Bernstein et al 2014)

Allowed foods and ingredients*

Soy-based infant formulas containing soy protein isolate, amino acid-based elemental infant formulas
All fruits, vegetables and their juices, pickled fruit and vegetables
All legumes (e.g., navy beans, kidney beans, garbanzo beans/chick peas, soybeans)
Soy-based products that are not fermented (soy milk, tofu, textured soy protein, hydrolyzed vegetable protein, soy protein concentrate, meat analogs)
Aged cheeses¹: Jarlsberg, Emmentaler, Swiss, Gruyere, Tilsiter, mature Parmesan, mature Cheddar cheese
Sodium and calcium caseinate
All cacao products except milk chocolate
Eggs
Additional ingredients: natural and artificial flavorings, all gums, including carrageenan

Foods used in moderation *

Soy sauce, soy products that are fermented (e.g., miso, natto, tempeh, sufu)
Meat by-products
Offal

Restricted foods and ingredients*

Breast milk, all milk-based infant formulas
Processed meats using lactose
All milk-based foods and beverages, including low lactose milk, except for caseinates and aged cheeses, listed above
All milk-based ingredients including buttermilk solids, casein, dry milk protein, dry milk solids, hydrolyzed whey protein, hydrolyzed casein protein, lactose, lactalbumin, whey
All cheese and cheese-based products except those listed above
Butter

¹ Galactose content and consequently allowed types of cheese may vary in different countries

* All manufactured foods need to be checked for the presence of milk by reading food ingredient labels

Foods used in moderation *

Soy sauce, soy products that are fermented (e.g., miso, natto, tempeh, sufu)
Meat by-products
Offal

Offal:

- its galactose content is unknown
- there is no direct evidence of harm
- It is a theoretical risk only



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Allowed foods and ingredients*

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- Soy-based products that are not fermented (soy milk, tofu, textured soy protein, hydrolyzed vegetable protein, soy protein concentrate, meat analogs)
- Aged cheeses¹: Jarlsberg, Emmentaler, Swiss, Gruyere, Tilsiter, mature Parmesan, mature Cheddar cheese
- Sodium and calcium caseinate
- All cacao products except milk chocolate

Eggs

Additional ingredients: natural and artificial flavorings, all gums, including carrageenan

Foods used in moderation *

- Soy sauce, soy products that are fermented (e.g., miso, natto, tempeh, sufu)
- Meat by-products
- Offal

Restricted foods and ingredients*

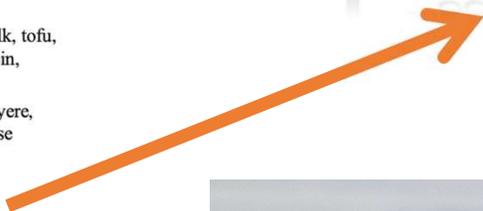
- Breast milk, all milk-based infant formulas
- Processed meats using lactose
- All milk-based foods and beverages, including low lactose milk, except for caseinates and aged cheeses, listed above
- All milk-based ingredients including buttermilk solids, casein, dry milk protein, dry milk solids, hydrolyzed whey protein, hydrolyzed casein protein, lactose, lactalbumin, whey
- All cheese and cheese-based products except those listed above
- Butter

¹ Galactose content and consequently allowed types of cheese may vary in different countries

* All manufactured foods need to be checked for the presence of milk by reading food ingredient labels

All cacao products except milk chocolate

Eggs



Dark chocolate



Calcium and Vitamin D

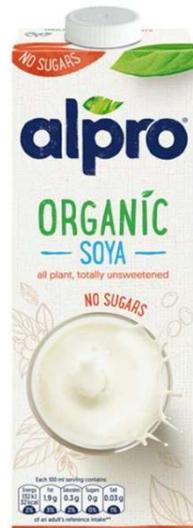
Recommendation #7 (+)

We recommend an annual dietary assessment of calcium and vitamin D intake with measurement of plasma total 25-OH-vitamin D levels. Both calcium and vitamin D should be supplemented as necessary following the age-specific recommendations for the general population.

Poll 9: Which has the highest amount of Calcium in it?



1) 200mls of a fortified soya drink



2) 200mls of a organic soya drink



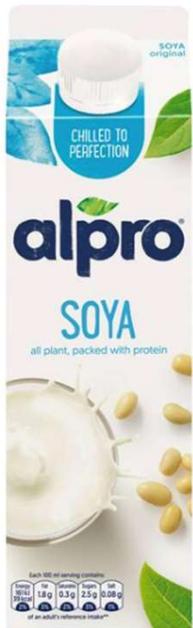
4) 80g of cooked spinach

3) 80g Cooked Kale



5) 30g of Emmental cheese

Poll 9: Which has the highest amount of Calcium in it?



1) 200mls of a fortified soya drink = 240mg



2) 200mls of a organic soya drink = 0mg



4) 80g of cooked spinach = 120mg but body can't absorb the calcium very well

3) 80g Cooked Kale = 120mg



5) 30g of Emmental cheese = 220mg

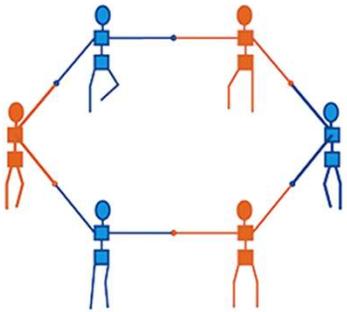
Poll 10: Which foods that are now allowed in the diet do you enjoy the most?

- 1) Fruit and vegetables
- 2) Legumes
- 3) Matured cheese
- 4) Soy based products

Conclusion

- The diet is just a restriction of dairy foods / foods containing milk (unless a suitable cheese, butter oil, milk fat or caseinates)
- The guidelines were the first attempt to make an International consensus of the diet
- They will be reviewed again and more evidence looked at

Where to find the guidelines



GalNet

The Galactosemia Network

 **Galactosemia guidelines summary
Bulgarian**

PDF | 326.68 KB

 **Galactosemia guidelines summary
Czech**

PDF | 328.79 KB

 **Galactosemia guidelines summary
Estonia**

PDF | 1.52 MB

 **Galactosemia guidelines summary
German**

PDF | 495.86 KB

 **Galactosemia guidelines summary
Portuguese**

PDF | 393.84 KB

 **Galactosemia guidelines summary
Croatian**

PDF | 253.84 KB

 **Galactosemia guidelines summary
Dutch**

PDF | 603.66 KB

 **Galactosemia guidelines summary
French**

PDF | 633.1 KB

 **Galactosemia guidelines summary
Italian**

PDF | 143.93 KB

 **Galactosemia guidelines summary
Spanish**

PDF | 979.17 KB

<https://www.galactosemianetwork.org/guidelines>

GalNet Nutritionist Representatives

Focus areas

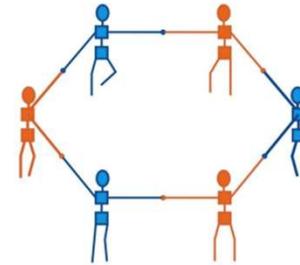
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Contact

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Q&A

- You can ask your questions now via the Q&A button
- Move your mouse to get the Zoom menu bar at the bottom of your screen
- The panelists will try to answer your question or combine with other questions
- If time runs short, we may have to 'dismiss' your question
- If your question wasn't answered, you can send your question to chairman@galactosaemia.eu

