



Galactosemia at a Glance

Galactosemia is a metabolic disorder, meaning the chemical process for breaking down food is not working properly.

Children with galactosemia are missing an enzyme that converts galactose into glucose for energy. Galactose is a simple sugar that is found in many foods, particularly dairy products.

One in 60,000 children is diagnosed with classic galactosemia in the United States. It is typically diagnosed within the first few weeks of life through a blood sample taken as part of a newborn screening. The child must be put on a strict dairy-free and galactose-free diet. This will help prevent severe complications such as cataracts, sepsis, multi-organ failure, brain damage or death. Maintaining this galactose-free diet is the only available treatment at this time. An international group called The Galactosemia Network recently reached agreement on a set of guidelines for dietary treatment for Classic Galactosemia. (<http://link.springer.com/article/10.1007/s10545-016-9990-5>)

The main points of the diet recommendations are listed below:

- Life- long galactose-restricted diet that eliminates sources of lactose and galactose from dairy products
- Galactose from non-milk sources that contribute minimal dietary galactose is permitted
- Fruits, vegetables, legumes, unfermented soy based products, mature cheese (see list below), and food additives sodium and calcium caseinate are allowed
- Annual dietary measurement of calcium or vitamin D is recommended and supplementation is recommended if necessary

Research suggests that despite strict dietary adherence, development, such as speech and motor, can be affected as early as pre-school age and continue into adulthood. Depending on how they are affected, children may have difficulties with communication, comprehension of new concepts, and sensory-motor integration skills.

NOTE: The information on these pages applies only to classical galactosemia. There are mild forms that may be called galactosemia variants that are not included in this section.

1. Medical / Dietary Needs

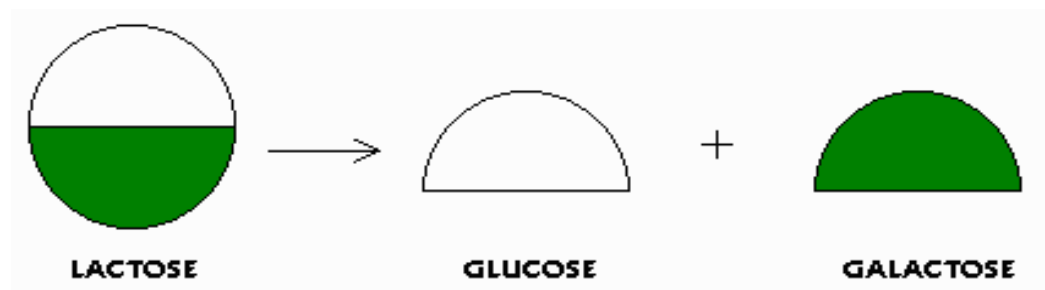
What you need to know

Galactosemia does not present the same in all children. Many have varying degrees of symptoms, and some do not show difficulties. If a newborn is untreated, he/she may have vomiting, diarrhea, and fail to gain weight.

Be aware, or ask a parent, if the child has a medical alert bracelet.

Dietary

Galactosemia affects the body's ability to process some sugars from food. Galactose is a sugar that may be found alone in foods but is usually found as part of another sugar called lactose.



Lactose is sometimes called milk sugar and is made of equal parts of two simple sugars: galactose and glucose. (source: A Teachers Guide to Galactosemia, Texas Dept. of Health, <https://www.dshs.state.tx.us/newborn/teachgal.shtm>).

People with galactosemia need to completely avoid foods with **galactose** and/or **lactose (glucose and galactose)**. This includes all dairy products from animals and foods with dairy products. If galactosemia is left untreated, galactose will accumulate in the blood and body tissues and will cause damage.

Children with galactosemia may need a 504 plan to accommodate dietary needs. A child may need special arrangements for lunch during the school day.

Allowed foods

- Fruits
- Vegetables
- Legumes
- Unfermented soy-based products

- Mature cheese (Jarlsberg, Gruyere, Emmentaler, Swiss, Tilster, grated 100% Parmesan, Parmesan aged >10 months, and sharp Cheddar cheese)
- Food additives sodium or calcium caseinate

Unacceptable foods

Food labels are our main tool for determining if a food or beverage is acceptable (or not) for the diet for galactosemia. Reading labels is key to eliminating as much galactose as possible.

Food Ingredients which are unacceptable in the diet for Classical Galactosemia:

- Butter
- Buttermilk
- Buttermilk Solids
- Cheese (EXCEPTIONS: Jarlsberg, Gruyere, Emmentaler, Swiss, Tilster, grated 100% Parmesan, Parmesan aged >10 months, and sharp Cheddar cheese)
- Cream
- Dough Conditioners*
- Dry Milk
- Dry Milk Protein
- Dry Milk Solids
- Ghee
- Hydrolyzed Whey**
- Ice Cream
- Lactalbumin
- Lactose
- Lactoglobulin
- Lactostearin
- Margarine***
- Milk
- Milk Chocolate
- Milk Solids
- Milk Derivatives
- MSG (Monosodium Glutamate)****
- Nonfat Milk
- Nonfat Dry Milk
- Nonfat Dry Milk Solids
- Organ Meats (liver, heart, kidney, brains, pancreas)
- Sherbet
- Sour Cream
- Fermented Soy products and Soy Sauce*****
- Whey and Whey Solids
- Yogurt

- Tragacanth Gum

NOTE: Lactate, Lactic acid and Lactylate do not contain lactose and are acceptable ingredients.

* Dough Conditioners may include caseinates which are UNACCEPTABLE. Most labels specify the name of the conditioner which is added to the product. If not, contact the company to make sure that all are acceptable.

** Hydrolyzed protein is UNACCEPTABLE and is commonly found in canned meats, like tuna. Hydrolyzed vegetable protein, however, is acceptable.

*** A few diet margarines do not contain milk. Check labels before using any brand. If "margarine" is listed as an ingredient in any processed food, consider the product UNACCEPTABLE.

**** MSG or Monosodium Glutamate itself is acceptable; however, some MSG's contain lactose extenders. It is best to avoid MSG whenever possible.

***** Soy sauce is UNACCEPTABLE if it is fermented. Brands must be checked before including this in the Galactosemic diet.

Taken from the Galactosemia Foundation / Diet Resources
(<http://www.galactosemia.org/diet-resources/>)

Medical

An individual with galactosemia may or may not experience any of the following potential complications.

- Cataracts (30%)
 - A cataract is a clouding of the lens of the eye.
 - In this population, cataracts are often mild, transient, and resolve with dietary treatment. They may form in one or both eyes and growth rate varies.
 - In general, it is believed that if a galactose restrictive diet is followed, cataracts should not develop.
- Learning difficulties
 - Speech/language difficulties (60%)
 - Problems range from mild to moderate or severe.
 - Individuals may have delayed vocabulary.

- A common type of speech difficulty found in individuals with galactosemia is apraxia of speech
 - This is often referred to as dyspraxia.
 - This is a motor speech disorder.
 - Difficulties with math or reading in school.
- Motor difficulties
 - Fine and/or motor difficulties may be present.
- Neurological (13-20%)
 - Ataxia balance, gait, and fine motor tremors
- Primary Ovarian Insufficiency: (POI)
 - Most females will exhibit POI
 - May have delayed menstruation
- Growth delayed (30%)
 - Growth may be severely delayed during childhood and early adolescence when puberty is delayed.
 - Growth continues through late teens.

What you can do

- Good communication with parents is very important.
 - *Let parent know if child has eaten any food not allowed*
 - If a special event (party, birthday) is coming up so a galactose-free food can be provided. Or have families send in a treat that can be stored in the classroom for these occasions.
- Little tastes can add up and shouldn't happen. Supervision of younger children with galactosemia may be needed to prevent sharing or "tastes."
- If you are unsure, do not give the food.
- School staff should treat a child with galactosemia as a normal healthy member of the class.
- Work with cafeteria staff to support the special diet and make it easy for the child to be included.
- Lactate, Lactic acid, Lactylate do not contain lactose and are acceptable ingredients.

Explaining dietary differences to classmates can be helpful. It is a good idea to involve the family and child in the explanation. A few ideas to think about:

- Children understand the idea of a food allergy.

- Discuss general differences within the class. Emphasize that all people are different.
- People eat different foods for various reasons (food customs, religious reasons, and regional differences, vegetarian, etc.).
- People have different diets (diabetes, etc.) to help their bodies.
- Involve the school nurse.

2. Education Supports

It is important to have **HIGH LEARNING EXPECTATIONS** for children who have **galactosemia**. Encourage use of the core educational curriculum and modify it in order to meet the individual needs of the child.

What you need to know

Work collaboratively on reinforcing successful strategies in the classroom as well as at home.

In the classroom, ***some or all*** of the following challenges may be observed.

- Difficulty communicating:
 - May ramble incoherently and erratically, but child may assume he/she is being understood (speech apraxia).
- Difficulty comprehending:
 - May stare blankly
 - Abstract concepts and opposites may not be understood.
- Poor motor planning/processing
- Poor sensory-motor integration
- Struggles holding a pen and with writing
- Becomes easily frustrated
- Difficulty remembering
- Lack of energy
- Impulsivity
- Desire to please and participate
- Motivated by praise

Parents may not want their child to be treated differently in school. School staff should try and be sensitive to this issue and convey a sense of normalcy. Parents may also experience their child's frustration and may feel frustrated themselves. In order to keep

strategies at home and school consistent, parents can relay to school staff what they have observed at home and what strategies they use.

What you can do

- Maintain a strict dairy-free and galactose-free diet
- Maintain a similar classroom structure/schedule

Communication strategies:

- Pace – break topics down slowly
- Pitch – change the tone of your voice when describing opposite concepts
- Pictures – use a visual aid, pictures, or sign language when possible. Provide a word bank if applicable
- Praise – provide positive feedback
- Patience – maintain patience. Present material in a slow sequential framework

Lesson plans:

- Packets – provide note packets for the week and distribute them prior to lessons
- Peers/Partners – place students next to peers or assign partners so they can emulate their behavior. One-on-one instruction if necessary.
- Prioritize – provide subject folders for assignments and an outline of expectations/goals
- Prompt – use preparatory teaching : tell the students when their turn is coming up

There are technological innovations that can be used in the classroom to help children with galactosemia progress along with other general education students. One example is the Kurzweil Educational System, Inc.:

- A computer system created for those with learning delays that adapts lesson plans based on the student's needs
- Documents can be scanned into the program and modified for the particular delay or difficulty. Students have the ability to hear what they write via headphones and can work on the same assignment as general education students simultaneously.

3. Behavioral and Sensory Support

What you need to know

Depending on needs of the individual child, therapies including physical, speech, and occupational, have been shown to increase the child's motor planning, processing, and integration skills.

What you can do

Some recommendations from therapists include:

- Breathing – advise the child to take a deep breath if speech is erratic
- Tracing – Allow the child to trace letters repeatedly
- Repetition – repeat exercises several times to enhance muscle memory
- Functional Training – practice exercises that mimic everyday movements
- Modeling – place the children next to peers so that they can model behavior
- Sensory Table – use of sensory table to work with different mediums

The classroom environment can also affect the child's progress.

- Structure – maintain a similar classroom structure and schedule
- Sequence – transition subject topics in the same pattern each day
- Instruction – one-on-one instruction with significant repetition may be very helpful

Look for patterns/behaviors in the classroom that can be reinforced in the home.

Have an open communication policy with parents, other clinicians and educational professionals.

4. Physical Activity, Trips, Events

What you need to know

The child may need to bring special foods on a trip. For special functions, a supply of allowable foods is good to have on hand. Preparation for trips may include foods and other supports.

If you live in New England (USA) and qualify, Northeast Passage offers Therapeutic Recreation and Adaptive Sports programming (www.nepassage.org).

What you can do

- Make sure parents are involved in planning for trips and functions.
- Volunteers need to be aware of dietary and processing issues.
- Supervision around any new eating situations needs to be arranged.

5. School Absences and Fatigue

What you need to know

It is important to have metabolic control throughout the lifetime to reduce risks associated with galactosemia.

What you can do

- Contact parents if any change is noted.
- Individuals with galactosemia should not need accommodations for fatigability.
- Individuals with galactosemia should not have increased absences due to their condition.

6. Emergency Planning

What you need to know

If a child accidentally has a food with galactose, it is important to let the parents know and to create a plan that might prevent that in future.

7. Resources

New England Consortium of Metabolic Programs

www.newenglandconsortium.org

This site has a wealth of information on metabolic conditions including galactosemia.

Understanding Galactosemia – an introductory guide for educators, featuring a basic description of galactosemia, suggested teaching strategies, classroom management ideas, and additional resources.

Galactosemia: Resources for Educators – a detailed guide for educators with in-depth information about galactosemia, about special challenges for students with the condition, and featuring information about therapies, classroom environment, lesson planning, teaching technologies, and other resources.

GSeeME – a website for educators with students who have galactosemia. The site includes a description of galactosemia, details about children’s special dietary needs, teachers’ classroom strategies, parent-teacher communication ideas, and other resources.

Galactosemia Foundation

Previously known as “Parents of Galactosemic Children Inc.”, their goal is to educate, support, and provide advocacy for those individuals affected by galactosemia.

<http://galactosemia.org/>

Understanding Galactosemia - A Diet Guide (PDF)

This site provides information about galactosemia as well as information to manage the diet, galactose content in foods and tools for meal planning.

<http://galactosemia.org/PDFs/UnderstandingGalactosemiaDietGuide3.pdf>

Texas Department of State Health Services

Offers a Galactosemia food card with "ok" foods and other dietary information for individuals with galactosemia.

<http://www.dshs.state.tx.us/newborn/>

Genetic Home Reference

Consumer-friendly information about human genetics from the U.S. National Library of Medicine

<http://ghr.nlm.nih.gov/condition/galactosemia>

GeneReviews

Learn even more about the genetics of Galactosemia at NCBI Bookshelf / GeneReviews.

<http://www.ncbi.nlm.nih.gov/books/NBK1518>