

# Galactosemia

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## Disclaimer:

This is not intended to be medical or clinical advice. Any recommendations here should be reviewed for the appropriateness for your individual child and their unique situation. Also, please note that each document should include the date that it was last reviewed or updated. Research and guidelines that have been released after that date may represent more up-to-date recommendations.

# Galactosemia

## At a Glance

*NOTE: The information on these pages applies only to classical galactosemia.*

Galactosemia is a rare genetic condition. Children with galactosemia are missing a chemical that breaks down *galactose*. Galactose is a simple sugar that is found in many foods, particularly dairy products.

If a child with galactosemia is given milk, substances build up in their tissues and cause damage. They must avoid dairy and be careful about eating other foods containing galactose. Even with a very strict diet, development can be affected in children with galactosemia. You may not notice anything different about a child with galactosemia since they won't look any different, but there are a few important characteristics to be aware of:

### Common Features of Galactosemia:

- Language/speech difficulties
- Difficulties with math or reading
- Problems with balance or walking
- Tremor or fine motor difficulties
- Behavioral issues
- Growth delay
- Delayed menstruation in girls
- Cataracts - clouding of the lens of the eye.

## In Depth

### Medical and Dietary Considerations

#### 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 any difficulties. Discuss with the parents the specific needs of their child. Because galactosemia is usually part of newborn screening, infants with this disorder are typically detected and treated very early in life, and the most serious complications can usually be avoided.

*Lactose* (milk sugar) breaks down to *galactose* so people with galactosemia need to completely avoid dairy products with galactose and/or lactose for their whole lives. If not, galactose will accumulate in the brain and other body tissues and will cause damage. Even with strict galactose avoidance, some children may have developmental issues

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 cheeses (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 the main tool for determining if a food or beverage is acceptable for the diet for galactosemia. Reading labels is key to eliminating as much galactose as possible from the diet.

Food Ingredients which are unacceptable in the diet for classical galactosemia include:

- Butter
- Buttermilk
- Buttermilk Solids
- Cheese (*except for those listed above*)
- 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 Complications

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

- Cataracts - often mild, resolve with dietary treatment
- Learning difficulties
  - Speech/language difficulties
    - Problems range from mild to moderate or severe
    - Individuals may have delayed vocabulary
    - Often this is a motor speech disorder - apraxia of speech or dyspraxia
  - Difficulties with math or reading in school
- Motor difficulties
  - Fine and/or motor difficulties may be present
- Neurological
  - Uncoordinated movements, balance problems, unsteady gait, tremors
- Primary Ovarian Insufficiency: (POI)
  - Most females will exhibit POI
  - May have delayed menstruation
- Growth delay
  - Growth may be severely delayed during childhood and early adolescence when puberty is delayed
  - Growth continues through late teens

## What you can do

Experts agree on the following dietary recommendations:

- 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 above), 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

Tips to maintain the strict diet

- Communicate openly with parents
  - Let parents know if their child has eaten any food not allowed
  - If a special event (party, birthday) is coming up, see if a lactose/galactose-free alternative can be provided. Parents may wish to send in a treat that can be stored in the classroom for these occasions.
- Avoid allowing 'little tastes' which can add up and be harmful. Supervision of younger

children with galactosemia may be needed to prevent sharing.

- If you are unsure, do not allow the food.
- Work with cafeteria staff to support the special diet and make it easy for the child to be included.

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 stay healthy
- Involve the school nurse

Except for dietary considerations, treat a child with galactosemia as a normal healthy member of the class.

## Education Supports

It is important to have high learning expectations for children who have galactosemia, utilizing their strengths and interests as a foundation. Encourage use of the core educational curriculum and modify it to meet the child's individual needs.

## What you need to know

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

- Difficulty communicating:
  - Inability to coordinate the complex oral movements needed to form syllables into words, words into phrases, and phrases into sentences (speech apraxia)
  - The child may not be aware they cannot be understood
- Difficulty comprehending:
  - May stare blankly
  - Abstract concepts and opposites may not be understood
- Poor motor planning/processing
- Poor sensory-motor integration
- Difficulty holding a pen and with writing
- Easily frustrated
- Difficulty remembering
- Lack of energy
- Impulsivity
- Desire to please and participate
- Motivated by praise

## What you can do

Work collaboratively on reinforcing successful strategies in the classroom. Encourage open communication with parents concerning frustrations seen at school and home.

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

Communication strategies:

- Break topics down slowly
- Change the tone of your voice when describing opposite concepts
- Use a visual aid, pictures, or sign language when possible. Provide a word bank if applicable
- Provide positive feedback
- Present material in a slow sequential framework

Lesson plans:

- Provide note packets for the week and distribute them prior to lessons
- Place students next to peers or assign partners so they can emulate their behavior. One-on-one instruction if necessary.
- Provide subject folders for assignments and an outline of expectations/goals
- Prompt students when their turn is coming up

## Behavioral and Sensory Support

### What you need to know

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

### What you can do

Have an open communication policy with parents, other clinicians and education professionals. Some recommendations from therapists include:

- Advise the child to take a deep breath if speech is erratic
- Allow the child to trace letters repeatedly
- Repeat exercises several times to enhance muscle memory
- Practice exercises that mimic everyday movements
- Place the children next to peers so that they can model behavior
- Use of sensory table to work with different mediums

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

- Maintain a similar classroom structure and schedule when possible
- Transition subject topics in the same pattern each day

- 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

## Physical Activity, Trips, Events

### What you need to know

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

### 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

## School Absences and Fatigue

Individuals with galactosemia should not experience fatigue more frequently than their classmates. They are unlikely to have increased absences due to their condition. Contact parents if any change is noted.

## Emergency Planning

Communicate with the parent to determine if there is a need to develop an emergency plan because of any medical issues. If a child accidentally has a food with galactose, it is important to let the parents know and create a plan to prevent that in future



## Resources

### **Medline Plus**

MedlinePlus is an online health information resource for patients and their families and friends. It is a service of the National Library of Medicine. This link will provide a good medical overview of the condition without being too technical.

<https://medlineplus.gov/genetics/condition/galactosemia/>

### **Galactosemia Foundation**

The Galactosemia Foundation is a nonprofit organization that advocates for individuals with galactosemia and their families. The website has a wealth of information. Check out the “Resources” tab for more in-depth information and helpful guidance tools.

<http://galactosemia.org/>

### **New England Consortium of Metabolic Programs**

The Consortium has created and collected a variety of valuable guidance tools for those with galactosemia. Explore those resources at this link

<https://www.newenglandconsortium.org/galactosemia-open-page>

## Info for School Nurse and Primary Care Staff

Galactosemia is a complex condition that will be best managed by good collaboration between the school nursing staff and primary care staff as part of the student's team. Work with the parents to get signed releases to share information at the school nurse's office and the primary care office.

*Gene Reviews* is a peer-reviewed point-of-care reference accessible via the National Library of Medicine. The entry for Galactosemia provides a comprehensive summary of the condition and describes some ongoing surveillance and management issues. It is a great resource for medical providers. <https://www.ncbi.nlm.nih.gov/books/NBK1518/>

### HIPAA Concerns

Sometimes concerns by primary care staff about violating HIPAA regulations can hamper care by limiting access to necessary medical information. Please note that if care is intended to be delivered by school staff, clarifying medical orders or medication instructions represents continuation of medical care and is not a violation of HIPAA. To avoid any issues like this, remind the parents to sign a release of information form at the primary care office. The school nurse should keep a copy of that release in the student's file, if possible, to facilitate sharing of information in the event that there is difficulty obtaining needed information.

### Example letter for physician to provide to school

"My patient \_\_\_\_\_ has been diagnosed with Galactosemia. This is a rare genetic condition in which the body is unable to process lactose or galactose. Strict dietary avoidance of those substances is essential. Children with Galactosemia are likely to have some growth delay and often experience developmental delays as well. There are variable medical complications that can occur. This condition necessitates an Individualized Education Program (IEP) or 504 plan.