

# **THE GALACTOSAEMIA HANDBOOK**

Prepared by the Australasian Society for Inborn Errors of Metabolism  
(ASIAM)

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## 2. Acknowledgements

Prepared by a Working Party of the Australasian Society for Inborn Errors of Metabolism, a special interest group of the Human Genetic Society of Australasia.

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### ACKNOWLEDGEMENTS

We would like to acknowledge and thank the many ASIEM members who contributed to the original handbook and provided constructive reviews of subsequent editions. We would like to note that ASIEM members are active members of the international galactosaemia clinical and research consortium:

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

Patients with galactosaemia are cared for in a multidisciplinary team environment and therefore, this edition of the handbook was reviewed by multidisciplinary metabolic teams across Australia and New Zealand prior to publishing. We would like to thank the various metabolic clinicians listed below who reviewed this handbook to ensure it is a complete and accurate resource for patients in Australasia.

- Dr Maureen Evans PhD, AdvAPD and Director of Metabolic Medicine, The Royal Children's Hospital Melbourne, VIC
- Professor Angela Morgan PhD, Speech Pathologist, Murdoch Children's Research Institute, Melbourne, VIC
- Professor David Coman, Metabolic Physician, Children's Health Queensland, QLD
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### INTRODUCTION TO THIRD EDITION

This Handbook replaces the second ASIEM Handbook for Galactosaemia published in 2010. The 2017 international guidelines for the management of classical galactosaemia (Welling et al., 2017) published since, support the inclusion of a wider range of foods than previously allowed in Australia and New Zealand. There have also been changes in food labelling within Australia and New Zealand, new recommendations on nutrient intake and additional data on the galactose content of foods. This information has been incorporated into this new edition.

## 3. About galactosaemia

### What is galactosaemia?

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Galactosaemia is an inherited condition that affects how the body processes a type of sugar called galactose. Galactose is an essential part of some structures within the body, and our bodies make some galactose themselves. Galactose is also part of lactose, the main sugar in animal milks (including breast milk, most infant formulas, cow's milk, goat's and sheep's milk).

There are several types of galactosaemia:

- The most common type is known as classical galactosaemia.
- Galactokinase deficiency (Rubio-Gozalbo et al., 2021) and galactose epimerase deficiency (Derks et al., 2022) affect different parts of the breakdown pathway of galactose and have different symptoms.
- Duarte galactosaemia is a form of galactosaemia which does not require treatment (Welling et al., 2017).

### What effects does galactosaemia have?

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Newborn babies with classical galactosaemia can become unwell in the first week of life and present with feeding difficulties, liver problems causing severe jaundice (yellow colouring/pigmentation of the skin) and bleeding problems, serious infections, and oil drop cataracts. Once they are fed with a galactose and lactose free formula, these problems improve rapidly. Often infants with galactosaemia require a short admission to hospital to treat their liver problems and set up their new diet. It is important to note that Galactosaemia is not a degenerative disease.

Treated children with classical galactosaemia grow normally, but there can be some problems during childhood and the adult years:

- About two thirds of children with classical galactosaemia have developmental difficulties, especially with language and/or speech (Panis et al., 2024 in Press). This is mostly delayed expressive language i.e. delay in spoken word developments but can also include issues with understanding and processing language (Panis et al., 2024 in Press). Speech therapy and learning support can be very helpful. The following link provides background information about normal speech development and helpful tips to support your child's speech development.

[Parenting SA - Learning to talk - Parent Easy Guide](#)

If you are concerned about your child's development, please talk to your metabolic team.

- Unfortunately, 90% of girls with classical galactosaemia will have ovarian function which affect progression to puberty and fertility problems (sometimes called ovarian failure or primary ovarian insufficiency) (Derks et al., 2023). Despite this, some women with galactosaemia have successfully had babies of their own.

Talk with your child's metabolic team to get the latest information about this issue. There are some tests (hormone level testing) which may be performed to check the condition of the ovaries in their pre-pubertal years. Slower than expected height growth pre-puberty can be a sign of future ovarian failure. If you are concerned, please discuss with your child's metabolic team.

*(Note: There is no evidence that galactosemia has any negative effect on the reproductive health of boys.)*

- Children with galactokinase deficiency are at risk of cataracts.
- Those with galactose epimerase deficiency have similar symptoms to classical galactosaemia.

## How is galactosaemia treated?

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### A GALACTOSE RESTRICTED DIET

At present there is no cure for galactosaemia. The treatment for galactosaemia is a lifelong diet excluding the major food sources of galactose. This is very successful in treating the symptoms in the newborn period. There is however still much to be learned about the long-term treatment of galactosaemia as good dietary control has not prevented the development of learning problems and ovarian failure in galactosaemia. It may be that some of the problems are caused before birth, or because of the body's own production of galactose.

The major food source of galactose is from lactose. Lactose is a sugar present in human and animal milks, which is broken down into glucose and galactose. This is why babies with galactosaemia must not be breastfed or fed standard infant formula and a special formula is needed. Lactose-free formula is not recommended as it still contains galactose.

Once solid foods are introduced, the foods will need to be milk and lactose free. Some foods contain obvious sources of milk, such as yoghurt, but other foods can contain hidden sources of milk or lactose added during manufacturing. For instance, some brands of canned spaghetti, sausage rolls, processed meats and snack foods may contain milk or lactose, and these brands should be avoided by a child with galactosaemia. The metabolic dietitian will teach you about the diet and help you manage it at different stages of your child's development.

By following a milk and lactose free diet it is possible to reduce galactose intake to less than 1-2% of the usual intake. There are some other foods that contain trace amounts of galactose, such as some fruits, vegetables, legumes and some animal products. However, international guidelines now recommend inclusion of all of these foods in the diet either in unlimited or moderate quantities (see section 6 for more detail). This is because the amount of galactose in these foods is much less than the galactose produced by the body itself.

Children with galactosaemia following these updated recommendations have not been found to have any adverse effects on long term health outcomes compared to a more restrictive diet.

### A MILK SUBSTITUTE

Appropriate milk substitutes are important in the diet to replace those nutrients usually provided by milk and milk products. If intake of the milk substitute is poor, it is important that your dietitian checks your child's diet for nutritional adequacy. Children and adults on milk free diets are particularly at risk of having a low calcium intake. Calcium is important for building strong bones and teeth and a low intake can increase the risk of fractures. An adequate calcium intake is essential from suitable foods or calcium supplements.

## How is galactosaemia diagnosed?

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Galactosaemia is diagnosed by the Newborn Screening Programme in most States of Australia and in New Zealand. This program tests the blood for many diseases and measures galactose-1-phosphate levels to test for galactosaemia. If necessary, the enzyme galactose-1-phosphate uridylyltransferase will also be measured.

Galactosaemia may also be suspected in a sick baby and the enzyme that is affected in galactosaemia is measured to confirm or exclude the diagnosis.

The metabolic clinic team – usually a dietitian, doctor, nurse practitioner, nurse, speech pathologist, genetic counsellor, laboratory scientist and a social worker or psychologist – provides ongoing support.

It is now possible to do expanded reproductive carrier screening to include the GALT gene which will indicate if you have an increased risk of having a child with galactosaemia prior to becoming pregnant. This is not currently covered by Medicare and therefore needs to be self-funded. <https://www.mackenziesmission.org.au>

## How did my child get galactosaemia?

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Galactosaemia is an inherited condition caused by faulty genes (see section 4 for more detail). It is considered a rare disease. Approximately 1 in 50 000 babies born in Australia and New Zealand have galactosaemia. This

means that only six or seven babies a year are diagnosed with Galactosaemia in Australia and New Zealand. People with galactosaemia are born with a deficiency of one of the enzymes needed to process galactose within the body.

### **Is galactosaemia the same as lactose intolerance or milk allergy?**

Galactosaemia is a genetic condition. It is not the same as lactose intolerance or milk allergy, which are far more common and are due to different causes. Milk allergy is due to a reaction by the immune system to the protein in milk. Lactose intolerance (or malabsorption) results from being unable to digest lactose effectively in the bowel. The diets for milk allergy and lactose intolerance are similar to the diet recommended for galactosaemia but are not suitable for a child with galactosaemia.

### **What about accidental exposures to galactose?**

Although the diet aims to be very low in galactose which is critical in the neonatal period and currently recommended for life, mistakes do happen by accident or misunderstanding. There is unlikely to be any immediate effect of this, except in the infant/young baby. If your child accidentally has small amounts of lactose, there are not expected to be any long-term effects.

## 4. What does the diagnosis of galactosaemia mean for my baby and our family?

### Your feelings as parents around the diagnosis

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If galactosaemia has been diagnosed because of symptoms of the disorder, you may be relieved that a diagnosis has been made. Even so, the information can still seem overwhelming.

If your baby has been diagnosed by the newborn screening test and seems well, accepting the diagnosis can be difficult. It is alarming to be told that this disorder can interfere with your child's health and development. The words used may be difficult to understand, and it may not be clear at first what the diagnosis really means.

The first few days or weeks can be stressful with a new baby. The feelings of joy, excitement and tiredness may be replaced with feelings such as disappointment, sadness and worry about your child's future. It is normal to feel worried about how you will manage. It is normal to have feelings of grief, disappointment, sadness or anger about what has happened. In these early stages, there is often a strong feeling of responsibility, and it is normal to worry or have doubts about how you will cope.

*"At first it was a bit daunting, but it got easier"*

The first concern a parent has for their newborn baby is his or her health and wellbeing. Many parents now choose to breast feed their babies and some women feel they have to breast feed their baby. Changing the way you feed your baby, suddenly from breast to a special formula may raise many questions regarding the health, development and the best care for your baby. The metabolic team looking after your baby will explain the changes needed for the best outcome and support you through these changes. As well as the physical, hormonal and emotional rollercoaster that this can be, you may feel concerned that your child will miss out on the benefits of breastfeeding. Some mothers may feel inadequate because they are not able to breast feed. These feelings are to be expected and talking with a health professional and/or supportive family members or friends may help.

You may also worry about your child's future, think about the things that will be different and what your child will miss out on. Most parents ask the question, "Why me?" or "Why my child?" at some point. Coming to terms with the diagnosis is a grieving process for many parents.

As you learn about the inheritance of galactosaemia, you may start to feel concerned about your family genes and guilty about passing the condition on. Having two people with the same gene for galactosaemia (GALT gene) is rare and has only recently been added to extended reproductive carrier screening. It can help to remind yourself that the diagnosis is not the result of anything you have done.

### About feeding your baby – important information for breast feeding mothers

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If you were breastfeeding your baby when galactosaemia was diagnosed, suddenly weaning your baby from the breast and introducing special formula may cause issues for you and will require advice and support to prevent discomfort. Seek help on comfortably reducing your breast milk supply from:

- your midwife
- a lactation counsellor / consultant
- your hospital's Maternal and Child Health Nurse or Breast Feeding Advisor
- your child and family health nurse or your Plunket nurse (New Zealand)
- Australian Breastfeeding Association (tel:1800 686 268)
- La Leche League

Australia: <https://www.llasia.org/lll-support-in-australia.html>

New Zealand: <https://lalecheleague.org.nz/>

## Donating expressed breast milk

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If you are thinking about donating your expressed breast milk, we recommend reading the following position statement and discussing with your midwife, obstetrician, or neonatologist further to ensure you are fully informed prior to making a decision.

Australia: [https://www.breastfeeding.asn.au/sites/default/files/2022-03/POL\\_StatementDonorMilk\\_V4.pdf](https://www.breastfeeding.asn.au/sites/default/files/2022-03/POL_StatementDonorMilk_V4.pdf)

New Zealand: <https://www.midwife.org.nz/wp-content/uploads/2019/06/Donor-Human-Milk-and-Milk-Sharing.pdf>

If your baby has been diagnosed as a newborn there is plenty of time to learn about the diet before your baby starts solid foods and you can take one step at a time.

## One step at a time

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Seeing an improvement in your child's health if he or she has been sick in the newborn period is reassuring and helps in the process of coming to terms with the diagnosis.

## Sharing the experience

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One thing that helps is to share your concerns and feelings with others. The early weeks and months are a time to allow trusted family members and friends to support you where possible. Your extended family and friends are always welcome to attend clinic appointments if you would find it helpful. Eventually a much wider circle of people will need to know. The Metabolic Clinic team will be able to put you in touch with support networks and organisations such as [Galactosemia Foundation \(https://galactosemia.org/\)](https://galactosemia.org/) and [Homepage | GalNet \(galactosemianetwork.org\)](http://galactosemianetwork.org).

## Explaining galactosaemia

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One of the challenges of having a child with galactosaemia is that, because it is rare, few people have heard of it. Finding ways of explaining the disorder as simply as possible to yourself, your family, your child, and interested others will evolve over time and as the need arises.

You may feel ready to tell family and friends about the diagnosis soon after it is made. But you may want to avoid telling too many people until you have adjusted to the diagnosis and the extra responsibility it brings. This allows you to let others know about the condition in your own time. Telling your relatives can be difficult, but it is important to do this early if you can, as ideally, the whole family needs to support each other.

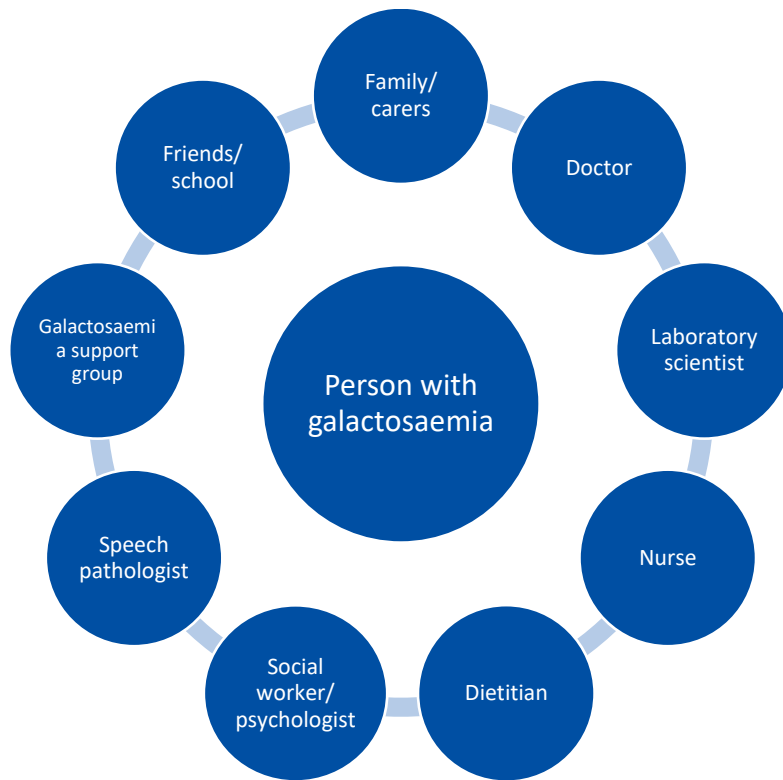
It will be important to talk about the diet, any special needs and what galactosaemia is with your child once they are at an age at which they can understand.

## A team approach

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Galactosaemia is not something parents can manage on their own. It is important to follow the instructions and recommendations of the metabolic team. Your child may also benefit from specialist help such as speech therapy.





*Figure 1: The galactosaemia support circle. You are not alone in managing your child's galactosaemia.*

## 5. Galactosaemia – the science behind the condition and the management

- Galactosaemia means galactose in the blood (aemia).
- Galactose from food and galactose produced by the body is either processed into compounds for essential structures within the body or is changed to glucose.
- Glucose is the sugar the body uses for energy.
- Changing galactose to glucose takes three main steps. Each step is driven by a special protein called an enzyme. In the three different types of galactosaemia one of these steps is affected. The faulty gene in galactosaemia causes the body to make inadequate amounts of the enzymes needed to breakdown galactose. The diagram below shows the breakdown of galactose in the body.

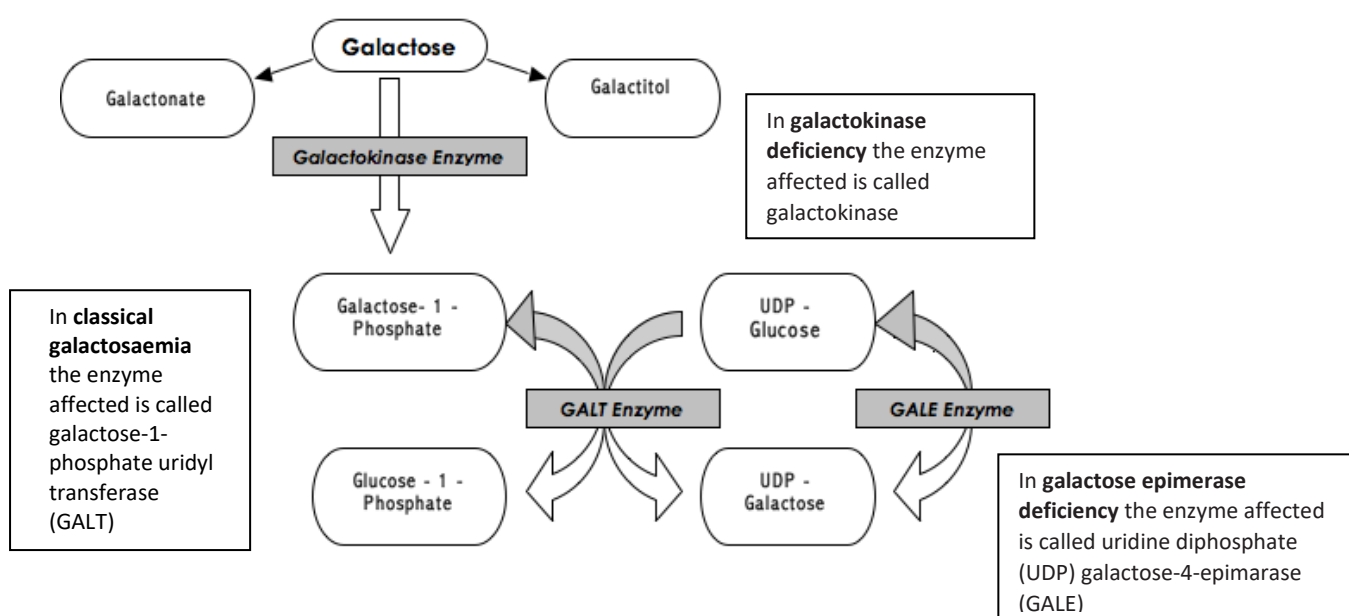


Figure 2: Galactose metabolism pathway.

This graphic was originally created by the authors of the first version of *The Galactosaemia Handbook* and permission was granted to re-use it in this version.

- Research is ongoing to improve the management of galactosaemia. ASIEM members, who are responsible for developing and updating this resource, are active clinical-research members of GalNet.
- Blood levels of galactose-1-phosphate (Gal-1-P) have limited use. They may detect major changes in diet, however levels do not seem to correlate with long term outcomes. Therefore, Gal-1-P is not typically monitored in Australia and New Zealand.
- UDP Galactose is used to make galactoproteins, galactolipids and mucopolysaccharides which are essential structural components of the body. It is thought that production of these important body components may be changed in galactosaemia.
- Galactose is also made in the body by reversal of the epimerase (GALE) reaction and from the natural turnover of galactoproteins and galactolipids. The body is therefore never 'galactose-free'. Production of galactose by the body is highly age dependent and varies between individuals.

## 6. Genetics and galactosaemia – frequently asked questions

### Will other family members be affected?

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Routine testing of other family members does not ordinarily occur, however if you would like to explore this, please ask your GP to refer you to your local genetics service. Any further babies you have will be tested soon after birth. There is a one in four chance that a full brother or sister of a child with galactosaemia will also have the disorder. Both boys and girls have the same chance of inheriting galactosaemia. Testing in pregnancy (antenatal testing) may be possible. There is no evidence that a galactose-restricted diet during pregnancy is beneficial to the outcome of a baby born with galactosaemia. It is best to discuss these issues with your metabolic doctor or a genetic counsellor prior to any new pregnancy.

### What is the role of genes?

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Galactosaemia is caused by a change to a gene also known as a genetic variant. Genes carry hereditary information about body processes and traits, such as blood group and hair colour, from parents to their children. Children inherit one set of genes from each parent. Arranged in pairs, these genes are replicated in every cell of the child and carry the 'blueprint' for each function and characteristic of their body. Children born with galactosaemia have inherited the same genetic variant from each parent.

### What happens when genes are faulty?

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Everyone carries some variations in their genes, usually without knowing which ones. In autosomal recessive disorders, a person who has one gene variant, but is not affected by it, is called a 'carrier' or 'genetic carrier'. Genetic conditions arise when two people with the same gene variant have children, as their child may be affected.

### How do faulty genes cause galactosaemia?

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The faulty gene in galactosaemia is one that controls the amount of the enzymes that break down galactose to glucose.

### What does having this faulty gene mean for my family?

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The pattern of galactosaemia inheritance is called 'autosomal recessive inheritance'. As described above, body cells carry two copies of each gene. However, the father's sperm cells and the mother's egg cells carry only one copy. For carriers of a single gene variant, each egg or sperm cell will carry either a gene with a variant in it or a gene without a variation in it. If, by chance, both parents carry one copy of the same gene variant, any baby conceived has:

- a one-in-four chance of inheriting two copies of the gene variation and is affected by the condition
- a two-in-four chance of becoming a healthy carrier like their parents
- a one-in-four chance of not carrying the gene variant at all

The diagram on the next page describes the inheritance of galactosaemia.

### If I have galactosaemia, will my children be born with galactosaemia?

If you have galactosaemia and your partner does not, the chance of having a baby with the condition is very low. For this to happen your partner would have to be a carrier and only about one in 150 people are likely to be carriers.

Testing for possible carrier status in a partner, from a family with no history of galactosaemia can be used to reduce the chance of having a child with galactosaemia, however a residual chance remains even if a person is not found to be a carrier. Prenatal testing is possible only if the gene variants in the family are known. Discuss testing with your metabolic doctor or a genetic counsellor.

### Inheritance of galactosaemia

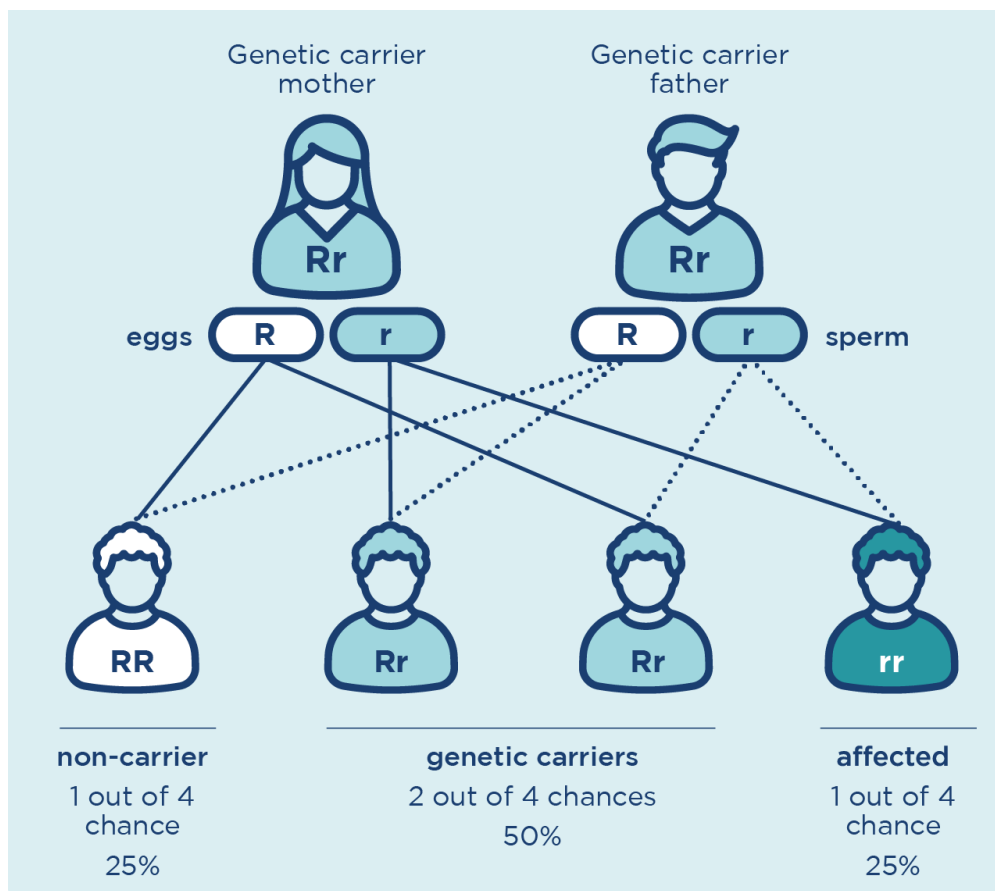


Figure 3: In autosomal recessive inheritance both parents are unaffected genetic carriers for the condition. The copy of the gene containing a recessive variant is represented by 'r'; the correct copy of the gene by 'R'. If both parents carry one galactosaemia gene variant there is a one in four chance that each of their children will have galactosaemia.

Reproduced with permission from Centre for Genetics Education Autosomal Recessive Inheritance Fact Sheet 7. Available from [Autosomal recessive inheritance \(genetics.edu.au\)](https://genetics.edu.au) (Accessed March 2024)

## 7. Feeding your baby with galactosaemia

### Overview

Babies with galactosaemia require a galactose free formula and are not able to be breast fed or to have standard infant formula, as both contain large amounts of lactose and galactose.

The formula recommended for your baby with galactosaemia will give them all of the nutrients they need during the first 6 months of life. Babies cope with the introduction of a bottle or a change of formula easily in the first few weeks of life. If you have any concerns with how your baby is feeding discuss this with your metabolic team.

Once you start your baby on solid foods you need to check these carefully to make sure they are milk and lactose free. Suitable foods for your baby are discussed in chapters 6 and 7 of this handbook.

### Choosing an infant formula

THE FOLLOWING INFANT FORMULAE ARE SUITABLE FOR YOUR BABY FROM BIRTH.

- Soy based infant formula
- Amino acid based formula or elemental formula
- Rice based formula

Your metabolic team will discuss specific products currently available at the time of your child's diagnosis

THE FOLLOWING INFANT FORMULAE ARE NOT SUITABLE FOR PEOPLE WITH GALACTOSAEMIA.

- Standard infant formula
- Other cow's milk based infant formula including:
  - Organic infant formula
  - Hypo-allergenic ("HA") formula
  - "Sensitive" or "comfort" or "gentle" formula
  - "Colic & Constipation" formula
  - "Reflux" formula
  - A2 milk formula
  - "Supreme", "gold", "expert", "pro" or "advance" formula
- Goats milk formula
- Lactose free ("LF") formula
- Partially or extensively hydrolysed formula

All of these formulae contain galactose (or may contain traces of galactose) and are not suitable for a baby with galactosaemia

## Preparing formula

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Refer to the link below for pictorial and video instructions.

[Infant formula preparation: in pictures | Raising Children Network](#)

[Baby formula, breastmilk & bottles | Raising Children Network](#)

Babies enjoy being held, cuddled and talked to during bottle feeds. This is important for their development.

Never leave your baby alone with a bottle or put your baby to bed with a bottle.

This link provides further information on giving your baby a bottle:

[Bottle-feeding babies: giving the bottle | Raising Children Network](#)

## How much formula should I offer my baby?

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It is best to feed your baby on demand – which means letting your baby decide when he/she is hungry and how much to drink at each feed. Don't force your baby to finish a bottle. If your baby starts turning his or her head away, fussing, or getting unsettled, they might be full.

Regular measurements of weight, length and head circumference are taken to make sure your baby is growing well. This can usually be done at your local health centre, and the results will be reviewed by the metabolic clinic team. In NZ, this is completed by your midwife (up to six weeks) then your Plunket nurse.

You will soon learn to recognise when your baby has drunk enough formula at a feed. Don't feel that your baby must finish every last drop of formula in a bottle.

If your baby is gaining weight as expected and has 6-8 wet nappies a day he/she is getting enough formula.

Talk to the metabolic team if you have any concerns about feeding your baby or feel you need help. Let them know if your baby is unwell or feeding poorly.

## How to clean and sterilise feeding equipment

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The following links provide information and pictorial instructions on cleaning and sterilising feeding equipment:

[Feeding Your Baby Infant Formula - HE1306 – HealthEd](#)

[Bottle-feeding: cleaning & sterilising | Raising Children Network](#)

## More information

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New Zealand has specific guidelines regarding use of tank and bore water, and these may be accessed on the following website:

[Feeding Your Baby Infant Formula - HE1306 – HealthEd](#)

## 8. Diet for galactosaemia

### Overview

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The diet for galactosaemia aims to exclude foods that are the major sources of galactose. The diet is for life.

The main food source of galactose is from lactose. Lactose is made up of glucose and galactose, and is the main sugar in milk and milk products. By following a lactose and milk free diet it is possible to reduce galactose intake to less than 1-2% of usual intake. Milk and milk products are also the main source of calcium in the diet and therefore ensuring adequate calcium intake from other sources is very important. Your dietitian will help you with this during clinic visits. See chapter 14 for more information.

There are some other foods that contain small amounts of galactose. Australian and New Zealand clinics do not exclude these foods in the diet for galactosaemia. In many cases it appears that the body cannot absorb this form of galactose.

The following pages give detailed information on which foods are allowed and how to check.

The diet for galactosaemia will be easier to manage if most of your meals and snacks are based on basic or home-made foods, rather than foods purchased away from home.

### Foods to avoid

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Current Australian and New Zealand recommendations are to avoid the following foods or ingredients:

- All animal milks - including human breast milk, cow's milk, goat milk, sheep milk, buffalo milk, camel milk and foods containing these milks (note that some hard cheeses are allowed).
- Lactose free milk, cream, yoghurt and custard (these still contain galactose).
- Lactose used in manufacturing of food and medicines.

There are a couple of other foods which contain small amounts of galactose which should be eaten in moderation. These include fermented soy products, by-products of meat/offal. See below for a full list.

## Galactosaemia diet checklist

The below checklist will help you to avoid lactose and galactose in the diet and offer alternative options that can be included in the diet instead. Please ensure you check all labels of packaged foods regardless of this checklist as ingredient lists may change more frequently than this handbook is updated.

Milk, milk products, and substitutes	
Allowed	Not allowed
<ul style="list-style-type: none"> <li>■ Soy based infant formula</li> <li>■ Rice based infant formula</li> <li>■ Other specialised formula prescribed by metabolic team</li> <li>■ Calcium fortified plant-based milk/ yoghurt/ ice-cream substitutes (<i>Soy, Rice, Oat, Almond and Coconut milk</i>)</li> </ul> <p>Notes:</p> <ol style="list-style-type: none"> <li>1. <i>Plant milks are nutritionally inadequate for infants under 12 months as a drink or formula replacement. They may be introduced in the preparation of food from 6 months and as a drink from 1 year</i></li> <li>2. <i>Do not use reduced fat varieties under 2 years of age</i></li> <li>3. <i>Should contain ~120mg calcium per 100ml</i></li> </ol>	<ul style="list-style-type: none"> <li>■ Breast milk</li> <li>■ Cow's/goat's/sheep milk based infant formula</li> <li>■ Lactose-free/lactose intolerance/low lactose formula</li> <li>■ Other infant formula not prescribed by Metabolic Team</li> <li>■ Cow's, goat's, sheep, camel milk (<i>fresh, UHT, powdered, evaporated, condensed</i>)</li> <li>■ Reduced or low lactose or lactose free milk, cream, yoghurt</li> <li>■ Fermented milk-based drinks</li> <li>■ Home-made plant-based milk substitutes as they do not contain any calcium</li> </ul>

Cheese		
Allowed	Include in moderation	Not allowed
<ul style="list-style-type: none"> <li>■ Vegan cheese including soy</li> </ul> <p>Note: <i>some soy cheese contains whey or casein which may contain some lactose. Check ingredients.</i></p>	<p>Generally, the harder and older the cheese, the lower the galactose content. The following cheeses are likely to have low galactose content:</p> <ul style="list-style-type: none"> <li>■ Hard cheese (<i>mature parmesan, mature cheddar, Gruyere</i>)</li> <li>■ Semi-hard cheese</li> </ul>	<p>All other cheeses except those listed in the allowed and include in moderation columns</p>

\* We recommend the following maximum serving sizes of hard/aged cheeses:  
 Up to 8 years of age = 1-2 slices (20-40g) per day (equivalent to ½ - 1 serve of dairy)  
 9 years and over = 2-4 slices (40-80g) per day (equivalent to 1-2 serves of dairy)

Desserts		
Allowed	Include in moderation	Not allowed
<ul style="list-style-type: none"> <li>■ Desserts made from allowed ingredients</li> <li>■ Jelly</li> <li>■ Dairy free cakes, slices, pies, puddings</li> <li>■ Plant based ice-cream/ yoghurt/custard (<i>check label to ensure there are no milk containing ingredients</i>)</li> </ul>	<ul style="list-style-type: none"> <li>■ Soy or tofu based ice confections/yoghurt/desserts.</li> </ul> <p>Note: <i>soy yoghurt is a fermented product and may contain some galactose released from fermentation of the naturally occurring sugars in the soy. Allowed in small quantities that are typically used in the diet.</i></p>	<ul style="list-style-type: none"> <li>■ Cream</li> <li>■ Dairy desserts e.g. custard</li> <li>■ Fromage frais (e.g. <i>Frêche</i>)</li> <li>■ Ice cream/ice confections</li> <li>■ Dairy based yoghurt/ Yoghurt products</li> <li>■ Lactose free yoghurt/dessert made with cow's milk</li> </ul>



Breads, cereals, pasta, rice, cakes, and biscuits	
Allowed	Not allowed
<ul style="list-style-type: none"> <li>■ Flour made from wheat, rye, barley, rice, maize (corn), soy, arrowroot</li> <li>■ Bran, cornmeal, wheatgerm, oats, rolled oats, ground rice, semolina, tapioca, sago</li> <li>■ Pasta, noodles (plain), cous cous, rice, barley, cracked wheat, polenta</li> <li>■ Homemade biscuits and cakes from allowed ingredients</li> </ul>	<ul style="list-style-type: none"> <li>■ Any bread, cereals, pasta, rice, cakes and biscuits (e.g. croissants, chocolate biscuits), crackers, rusks containing milk or lactose</li> </ul>

Fruits, vegetables, and legumes		
Allowed	Include in moderation	Not allowed
<ul style="list-style-type: none"> <li>■ Fruit: fresh, dried, frozen</li> <li>■ Fruit juice, fruit juice drinks</li> <li>■ Vegetables: fresh, frozen, dried or canned in brine or water with no other ingredients</li> <li>■ Legumes: (dried peas, beans, lentils, chickpeas *)</li> <li>■ Chick pea flour, besan flour, chick pea dahl, hummus, falafel made with chick peas</li> <li>■ Unfermented soy-based products</li> </ul>	<ul style="list-style-type: none"> <li>■ Fermented soy products, miso, tempeh, natto, soy sauce** <i>(see chapter 11 for more information on fermented soy products)</i></li> </ul>	<ul style="list-style-type: none"> <li>■ Any fruit based desserts or pastries containing sources of galactose e.g. apple custard</li> <li>■ Fruit smoothies containing animal dairy or lactose-free milk</li> <li>■ Instant mashed potato</li> <li>■ Vegetables served with galactose containing ingredients, cheese sauce</li> </ul>

\* We recommend the following maximum serving sizes of legumes:

Up to 8 years of age = ½– ½ cup

9 years and over = 1 cup

\*\*We recommend the following maximum serving sizes of fermented soy products:

Tempeh = 2-3 slices (40-60g) per day

Miso = 1 cup miso soup or 1Tbsp miso paste per day

Natto = 1 small serve (40g) per day

Soy sauce = 1 Tbsp/day

Fats and oils	
Allowed	Not allowed
<ul style="list-style-type: none"> <li>■ Milk free margarine e.g. Becel, Nuttelex, Olivani</li> <li>■ Oil</li> <li>■ Copha, lard, dripping</li> <li>■ Ghee</li> </ul>	<ul style="list-style-type: none"> <li>■ Imitation cream</li> <li>■ Butter</li> <li>■ Regular margarine</li> <li>■ Cream</li> </ul>

Drinks	
Allowed	Not allowed
<ul style="list-style-type: none"> <li>■ Calcium fortified plant-based milk (<i>plain or flavoured with fruit or cocoa and sugar or other allowed milk flavourings – see products list</i>)</li> <li>■ Tea, coffee (<i>no milk</i>)</li> <li>■ Fruit juice, soft drinks, cordials, soda water, tonic water, mineral water</li> </ul>	<ul style="list-style-type: none"> <li>■ Milk, milk shakes, flavoured milk</li> <li>■ Milk flavourings containing lactose e.g. malted milk powder, hot chocolate powder</li> </ul>

Meat, fish, eggs, and poultry		
Allowed	Include in moderation	Not allowed
<ul style="list-style-type: none"> <li>■ Meat: plain fresh or frozen</li> <li>■ Poultry: plain fresh or frozen</li> <li>■ Fish: plain fresh, frozen, canned in oil</li> <li>■ Eggs: plain or with allowed ingredients</li> </ul>	<ul style="list-style-type: none"> <li>■ Offal (<i>liver, kidney, brain, sweetbreads and pancreas</i>) contain some galactose and should not be eaten in large amounts e.g. pâté ***</li> <li>■ Egg substitute, yolk free egg mix</li> </ul>	<ul style="list-style-type: none"> <li>■ Pre-prepared meat/meat alternatives containing sources of galactose</li> <li>■ Meats/meat alternatives in marinades/sauces containing sources of galactose</li> <li>■ Corned meat containing sources of galactose</li> <li>■ Sausage rolls/meat pies (<i>pastry usually contains butter</i>)</li> <li>■ Tinned meat/meat alternatives containing sources of galactose</li> <li>■ Deli meats containing sources of galactose (<i>salami, mortadella, sausages, burgers</i>)</li> <li>■ Seasoning mixes containing lactose</li> </ul>
<p><b>REMEMBER: Always check with your butcher, chicken, poultry shop or take away shop about ingredients in sausages and other ready-made products.</b></p>		

\*\*\*We recommend the following maximum serving sizes of offal products:

Pâté = 2 tsp (10g) per day

The kitchen cupboard		
Allowed	Include in moderation	Not allowed
<ul style="list-style-type: none"> <li>■ Sugar, honey, jam, marmalade, golden/maple syrup, molasses.</li> <li>■ Allowed Sweeteners: <i>Aspartame (Equal, Nutrasweet)</i> <i>Acesulfame K (Sweet One)</i> <i>Saccharin (Sweet 'N Low, Sugar Twin)</i> <i>Stevia</i> <i>Sucralose (Splenda)</i> <i>Sugar Alcohols (e.g. xylitol, mannitol)</i></li> <li>■ Vegemite, Marmite, Peanut butter</li> <li>■ Salt, pepper, vinegar, herbs, spices.</li> <li>■ Sweets/lollies with nil lactose containing ingredients e.g. boiled sweets, gummy lollies, jelly beans</li> <li>■ Milk-free or dairy –free chocolate</li> <li>■ Potato/corn/vegetable chips</li> <li>■ Nuts</li> <li>■ Coconut, coconut milk/cream</li> <li>■ Plain/salted popcorn</li> </ul> <p><i>Note: some flavourings will contain milk e.g. butter or cheese flavour – check the ingredients or make at home.</i></p>	<ul style="list-style-type: none"> <li>■ Miso, Tempeh and Natto</li> <li>■ Soy sauce **</li> </ul>	<ul style="list-style-type: none"> <li>■ Any foods with milk-based ingredients including buttermilk solids, casein, dry milk protein, dry milk solids, hydrolysed whey protein, hydrolysed casein protein, lactose, lactalbumin, whey</li> </ul>

\*\*We recommend the following maximum serving sizes of fermented soy products:

Tempeh = 2-3 slices (40-60g) per day

Miso = 1 cup miso soup or 1Tbsp miso paste per day

Natto = 1 small serve (40g) per day

Soy sauce = 1 Tbsp/day

## 9. Label reading

### Overview

In Australia and New Zealand, food manufacturers must identify ingredients that are sources of milk in a product by writing it in the ingredient panel. You should carefully read the ingredient list on any food packet prior to consuming it to ensure the food does not contain any sources of galactose. Milk and lactose can be called different names in the ingredient list, some of which are simple to identify, and others that are not so obvious. All food labels must have certain information on them. Here is what to look for when reading a label for someone with galactosaemia.

The nutrition information panel must have a table listing the energy and nutrients (protein, fat, carbohydrate and sugars) per serve and per 100g or 100ml.

If a pack has a claim about another nutrient such as calcium, then this must also be listed.

Note that milk is present in several different ingredients.

There are several common allergens that must be highlighted. Milk is an allergen, and this rule is useful for individuals with galactosaemia.

### Sweet Biscuits

#### Nutrition Information

Servings per pack: About 20

Average Serving Size: 25g

Average quantity:	Per serving	Per 100g
Energy	515kJ	2060kJ
Protein	1.2g	4.8g
Fat, Total	5.7g	22.8g
- Saturated	3.3g	13.2g
Carbohydrate	16.5g	65.8g
- Sugars	8.1g	32.2g
Sodium	104mg	416mg

#### Ingredients:

Wheat flour, sugar, vegetable oil (contains soy), butter (from milk), desiccated coconut, compounded chocolate (contains emulsifier E322 from soy), oats, golden syrup, chocolate (contains milk, emulsifier E322 from soy), raspberry flavoured filling, colour (E129, E132), eggs, salt, raising agent, condensed milk, milk solids, honey, emulsifier (E322 from soy), flavour.

**May contain traces of nut and sesame.**

Ingredients are listed in order of highest to lowest weight: wheat flour is the major ingredient followed by sugar, vegetable oil and butter.

The additive numbers are given so you can find out what they are. There are no additives that are sources of milk or lactose.

#### 'May Contain' Warnings.

'May contain' statements are useful for individuals with severe food allergies. There is no need to avoid products that state 'may contain milk' if milk and other sources of galactose are not listed in the main ingredients list. There is also no need to avoid a product if it is 'manufactured in a plant which handles milk' if there is no other source of galactose listed in the main ingredients list.

If a food does not have a label (e.g. bread purchased from a bakery, or take away foods), legislation states that information regarding the ingredients of a product must be 'provided to the purchaser upon request'. You can ask for an ingredients list if you are not sure about a product. If you are not sure the information given is correct it is wise to avoid the food.

## 10. Medications and nutritional supplements

Medications - particularly tablets and capsules - often contain lactose. Ask your doctor or pharmacist to help you choose a brand of medication, if possible, which is free of lactose.

Medicines that contain lactose should include the statement:

*"This product contains lactose"*

(Therapeutic Goods Act Order no 48).

Doctors or pharmacists can also check for suitable products.

All prescribed medicines are safe for galactosaemia when given for short periods or in an emergency. If you are only using the medicine for a few days and there is no easy alternative, use the prescribed medicine recommended by your doctor even if it contains lactose. If medication is needed for a longer time (more than 1-2 weeks) and an alternative without lactose is not available, discuss this with your metabolic team.

Many nutritional supplements are based on cow's milk – always carefully check the ingredients for sources of galactose before you use them.

Some digestive aids (for bloating or wind) may contain alpha galactosidase. This may break down otherwise indigestible sources of galactose and should be avoided.

# 11. Controversial foods

## Overview

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If you do your own reading about the diet for galactosaemia outside of what is included in this handbook, you will note that some countries restrict many foods that are not restricted in Australia and New Zealand. Some foods, particularly plant foods (fruits, vegetables, and legumes) contain small amounts of galactose and are restricted in the diet for galactosaemia in some countries. In Australia and New Zealand, current recommendations allow foods that contain very small amounts of galactose, to be included in the diet for galactosaemia. A comparison of blood levels of galactose-1-phosphate in children with galactosaemia in NSW and SA found no difference using the current recommendations compared with a more restricted diet. Research in America and Holland also supports these findings. This is probably because the body can make some galactose itself with estimates suggesting that adults produce 1000–2100 mg galactose each day. It may also be because the galactose is sometimes bound with other indigestible components and the body cannot absorb it as well. A less restricted diet is healthier, more enjoyable and reduces the risk of inadequate nutrient intakes during times of rapid growth and development.

## Cheese

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In some hard matured cheeses, almost all the lactose (and the galactose) is removed when the milk is separated to make the curd or is consumed by the bacteria culture used to make the cheese during the maturation process.

However, published data on the lactose and galactose content of cheese varies. This is because cheeses of the same name may be made by different processes, may contain different cultures and/or be matured for different length of time. Lactose and galactose levels can vary within the same batch of cheese.

Australian and New Zealand cheese tends to be made by traditional manufacturing processes and as a general rule, the harder and more matured the cheese is, the less galactose will be present. Hard matured or vintage cheddar is the best choice and is a good source of calcium. See chapter 8 for further information.

The cheeses most likely to contain significant amounts of galactose are cream cheese, cottage cheese, ricotta, feta, processed cheese, haloumi, and neufchatel. These should be avoided.

## Cocoa

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Cocoa may contain bound galactose but is considered acceptable for people with galactosaemia. Chocolate bars usually contain other lactose containing ingredients such as milk so make sure you remember to read the labels!

## Eggs

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Eggs contain a very small amount of galactose in a bound form. These can be included.

## Fermented foods (e.g. pickles, sauerkraut)

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The fermentation of these foods may release any bound galactose in the original product. The level of galactose is likely to be very small, so intake of these fermented foods is acceptable.

### **Gums and fibres (e.g. acacia, agar, carrageenan, carob, guar gum, gum Arabic, locust bean gum, tragacanth gum, xanthan gum)**

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These may contain some bound galactose. It is unclear how much is available to be absorbed. These can be included in the diet.

### **Legumes (eg beans, lentils and dried peas)**

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Research has suggested that some legumes contain higher amounts of free galactose as well as galactose in the bound form. These were therefore previously restricted in the diet for people with galactosaemia. However, the recent international guidelines have stated that the galactose content of legumes is still far less than what the body produces itself and therefore are acceptable to include in the diet in usual quantities. Research has shown no adverse effects of including these foods in the diet.

### **Organ meats (eg liver, kidney, brains, sweetbreads and pancreas)**

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The galactose content of meat by-products/offal is unknown. There is no direct risk of harm, but the international guidelines recommend these products are eaten in moderation only due to a theoretical risk of containing galactose. Note that pâté is made with liver and should be included in small amounts only.

### **Seeds and nuts**

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These may contain bound galactose but are acceptable in the diet.

### **Soy beans and soy products**

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Soy beans are acceptable in the diet along with tofu and soy drinks. It is recommended that fermented soy products like tempeh, natto are included in moderate amounts. See chapter 8 for more information. Galactose may be released from bound sources during the manufacturing of soy sauce, but small amounts will not contribute significantly to the galactose intake and therefore can also be included in moderation.

Soy yoghurts do not currently contain added lactose, but it is not known whether galactose is released from the bound forms naturally found in soy during the fermentation process.

### **Soy formula and soy drinks for infants and young children**

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The use of soy formula and soy drinks for infants and young children has become controversial. The concerns regarding soy are mainly related to potential effects of the naturally occurring soy phytoestrogens (isoflavones) on hormone function. Although studied by numerous investigators in various species, there is no conclusive evidence from animal, human adult, or infant populations that dietary soy isoflavones adversely affect development, reproduction, or hormonal function. In their position statement regarding the use of soy formula, the Australian College of Physicians states that based on current evidence, soy formulas are still recommended for infants with galactosaemia as total exclusion of dietary lactose is more important than the potential risk posed by phytoestrogens in this group of infants. The New Zealand Ministry of Health state that soy based infant formula is indicated for galactosaemia as first line treatment.

## 12. Recipe adaptation

### Tips and tricks

Collect recipes that are free of milk and milk products or adapt favourites using soy drink instead of cow's milk, and milk free margarine instead of butter. It is easy to modify your own recipes to make them lactose free and suitable for someone with galactosaemia. Do not use 'lite' or lower fat milk free margarine in baking as these have a higher water content and will affect the success of your final product.

Here are a few recipe hints that can be used and are suitable for the whole family.

If a recipe calls for:	Use:
Butter or margarine	Milk free margarine
Cow's milk	Soy drink, soy formula, rice or oat drink
Ricotta cheese or cottage cheese	Silken tofu may be pureed to use instead of ricotta cheese or crumbled to use instead of cottage cheese
Cream cheese	Soy based cream cheese (*check the label carefully to make sure that it does not contain cow's milk)
Yoghurt	Soy yoghurt
Buttermilk	Substitute 1 cup with ½ cup soy yoghurt and ½ cup soy or rice drink OR 1 cup soy drink mixed with 1 tsp vinegar
Evaporated Milk	Evaporated soy milk
Cheese	Suitable matured cheese (see chapter 8)
Ice Cream	Soy based ice cream, milk free ice blocks or milk free gelato
Custard	Use soy or rice drink and custard powder or see recipe below
Soups and Gravies	Use water or soy drink, not milk or cream. Coconut milk is good to use in some soups
Porridge	Make porridge with water or soy drink

### Some useful recipes

Please note, for the below recipes:

Tbs = Tablespoon    tsp = teaspoon

#### CUSTARD

*You can make custard using custard powder and soy drink, but this recipe is colour free and suitable for babies and young children.*

- 250ml soy formula or soy drink
- 1 Tbs sugar
- 2 Tbs cornflour
- 2 drops vanilla essence

Blend sugar and cornflour with a small amount of formula, then add the rest of the formula. Microwave 3 minutes on high. Stir, if custard is not thick enough, microwave for another minute. Add vanilla essence last.

### WHITE SAUCE

- 1 Tbs milk free margarine
- 2 Tbs plain flour
- 1 ¼ cups soy or rice drink

Melt the margarine in a saucepan and remove from heat. Add the flour and stir until the mixture is smooth. Return to a gentle heat and cook 1 minute, stirring well. Add the liquid gradually and stir until smooth. Return to heat and stir until boiling. Reduce heat and continue to stir for 1-2 minutes. Remove from heat.

### TUNA MORNAY

Using one quantity of the white sauce recipe as a base, add 1 can of tinned tuna, ½ cup of grated matured cheese and ½ cup cooked frozen peas and corn.

### PIKELETS

- 2 cups plain flour
- 1 egg
- 1 ½ cups soy or rice drink
- 75g melted milk free margarine
- 3 tsp baking powder
- ½ cup sugar

Sift the flour and baking powder. Add the sugar. Slowly add the egg, soy or rice drink and margarine, mixing constantly until the batter is smooth. Cook spoonfuls over medium heat in a non stick frying pan until bubbles surface. Flip and cook until golden.

### SCONES

- 2 cups self raising flour
- 1 Tbs milk free margarine
- ¾ cup soy or rice drink

Heat oven to 230°C. Grease oven tray. Sift flour. Rub in margarine with fingertips. Add soy or rice drink and mix to a soft dough. Turn onto a lightly floured board and knead until smooth. Roll out 1 ½ cm thick. Cut into shapes with a floured cutter. Place on oven tray. Glaze tops with soy or rice drink. Bake 7-10 minutes or until golden brown, and the sides of the scones are set.

### HONEY CRACKLES

- 75g milk free margarine
- ¼ cup sugar
- 2 Tbs honey
- 4 cups corn flakes

Melt margarine, sugar, and honey together. Pour margarine mixture over corn flakes and mix to combine. Place spoonfuls of mixtures into patty pans. Bake at 180°C for 10 minutes.



### CUP CAKES

- 60g milk free margarine
- ¼ cup sugar
- 1 egg
- 1 cup self raising flour
- 2 Tbs soy or rice drink
- ¼ tsp vanilla essence

Heat oven to 160°C. Grease patty tins or line with patty cake papers. Beat the margarine and sugar. Add the egg and beat well. Sift the flour and add alternately with the soy or rice drink, a third at a time, folding in lightly. Fold in the vanilla essence. Spoon into prepared patty cake tins. Bake until golden brown, approximately 15 minutes. Ice with basic soft icing made with ¾ cup icing sugar mixture and 1 Tbs water.

### BANANA CAKE

- 125g milk free margarine
- ¾ cup brown sugar
- 2 very ripe bananas, mashed
- 1 tsp baking soda
- 1 Tbs hot water
- 1½ cups flour
- 1½ tsp baking powder
- 1 tsp cinnamon
- ½ cup soy or rice drink

Beat together margarine and sugar until well mixed. Add mashed bananas and mix well. Mix baking soda with hot water and add to banana mixture. Stir in flour, baking powder, cinnamon and soy or rice drink and mix gently until just combined. Pour into greased and lined 20 cm cake tin. Bake at 180°C for 30-35 minutes. Ice with chocolate icing.

### SPICED APPLE CAKE

- 100g milk free margarine
- 1 cup brown sugar, firmly packed
- 2 large apples, grated
- ½ cup sultanas
- 1 tsp baking soda
- 1 Tbsp hot water
- 1 tsp mixed spice
- 1 tsp cinnamon
- 1 cup flour
- ½ cup rolled oats
- 2 tsp baking powder

Melt the margarine in a large saucepan or bowl in microwave. Add brown sugar, apples and sultanas and mix well. Dissolve baking soda in hot water and add to apple mixture. Add remaining ingredients and mix gently until just combined. Pour into a greased and paper-lined 22cm round cake tin. Bake at 180°C for 45-50 minutes. Drizzle with lemon or orange icing.

### CHOCOLATE CAKE

- ¾ cup sugar
- 1½ cups flour
- 1 tsp baking powder
- ½ tsp baking soda
- ½ tsp salt
- 3 Tbs cocoa
- 1 tsp vanilla essence
- 1 Tbs malt vinegar
- 5 Tbsp oil
- 1 cup lukewarm water

Sieve together the flour, baking powder, baking soda, salt, and cocoa. Add sugar and mix. Add the vanilla essence, malt vinegar, oil, and water. Mix until just combined. Bake at 180°C for approximately 25-30 minutes in a lined and greased 20cm square or round cake tin. Spread chocolate icing over cake once cooled.

## ICINGS

### *Chocolate Icing*

- 1 cup icing sugar
- 2 Tbs cocoa
- 3 - 4 Tbs warm water

Mix icing sugar, cocoa, and water together to make a smooth paste.

### *Lemon or Orange Icing*

- 1 Tbs milk free margarine
- Juice of ½ a lemon or orange
- Icing sugar

Mix icing sugar, margarine, and juice together to make a smooth paste.

## ANZAC BISCUITS

- 1 cup flour
- 1 tsp baking powder
- 1 cup coconut
- 1 cup brown sugar
- 1 cup rolled oats
- ½ tsp baking soda
- 2 Tbs hot water
- 125g milk free margarine
- 1 Tbs golden syrup

Mix dry ingredients together. Melt margarine and golden syrup. Dissolve baking soda in hot water. Add margarine, golden syrup and baking soda to dry ingredients and mix well. Roll into small balls, flatten with a fork, and place on greased / floured tray. Bake at 190°C for 10-15 minutes. When cooked, transfer to a wire cooling rack until cold.

## 13. Special considerations

### Starting your baby with galactosaemia on solids

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Other than sticking to the diet for galactosemia (see chapter 8), you can follow the same advice given to the general population. The below link provides helpful information for when, why, what, and how to introduce solids.

[Introducing solids: why, when, what & how | Raising Children Network](#)

### Feeding toddlers and children

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It is important to foster healthy eating habits in all toddlers and children, including those with galactosaemia. The following links provide useful information about healthy eating for toddlers and children and how to deal with fussy eating.

[Healthy eating habits for children | Raising Children Network](#)

[Fussy eating in children: tips to help | Raising Children Network](#)

#### REMEMBER THAT:

- As a parent you give your child healthy food and opportunities to eat it. Your child can decide how much to eat – or whether to eat at all
- It is quite normal for children's appetites to vary from one day to the next
- If they are growing well, they are eating enough
- When feeding toddlers, expect waste and mess
- The metabolic team is there to help if you are having trouble with feeding

### Help your child understand their special diet.

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A child with galactosaemia may find it difficult to understand why they can't always eat the same foods as others.

Although being on a diet can seem hard, it is more difficult for your child if he or she doesn't receive firm guidelines about the foods that can be eaten and not eaten. It is important not to give into tantrums about wanting a forbidden food - it will make it worse the next time. Say and mean "no" but offer alternative foods.

While it is easier to use some milk free foods such as margarine or bread for the whole family, making the house 'cow's milk free' does not teach your child that he or she does have to eat differently.

Make sure everyone in the house has an adequate calcium intake by serving mealtime milk drinks for the whole family (soy drink for your child with galactosaemia).

People who look after your child must also realise the importance of your child's diet. People tend to think a milk free diet is for an allergy or lactose intolerance. Some children who are said to be allergic or intolerant to milk, sometimes have small amounts of milk or milk containing foods. So it is important that the person looking after your child knows that no food containing milk, lactose or galactose can be eaten by your child.

Try to offer simple explanations to your child about their need for a special diet. These explanations will become more complex as they grow older. Ask your metabolic team or other parents about approaches they have found helpful.

## Eating away from home

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### FOOD AT CHILDCARE

Most child care centres will be familiar with children requiring a milk free diet because of food allergies, and many will be able to supply a suitable milk free menu. Talk to them about your child's diet and give them a copy of the information sheet (chapter 15). Some child care centres will ask you to supply food from home.

### FOOD AT SCHOOL

Sending a packed lunch and snacks is safer than relying on foods that are purchased from the school canteen. Choose foods your child enjoys and which are allowed in the diet for galactosaemia.

Because of the prevalence of food allergies, swapping foods at school is discouraged but it still happens. Talk to your child about how to handle food swaps in the playground. Your child should only swap foods they recognise as being allowed in their diet.

Check the school canteen menu and make sure your child knows which foods are suitable to buy. Unfortunately suppliers can change frequently, so it is best to keep to a simple list. If possible, take your turn at working at the canteen so that your child can have a lunch order that day.

Keep a supply of small cakes or muffins in the freezer for cake stall days and birthdays. Send soy drink for breakfasts and other alternatives for other theme days.

Give your child's teacher a copy of the information sheet about galactosaemia provided in this handbook. They might wish to keep a container of milk free biscuits or lollies in the classroom as a substitute for birthday cakes or other treats. Encourage your child's teacher to use non-food rewards such as stickers, pencils or erasers etc.

If cooking is planned in the classroom, find substitutes your child can make or send a note to the teacher reminding him or her why your child should not taste foods containing milk. At secondary school, food technology lessons can be modified so that your child can cook foods that are suitable for their own diet.

### GOING TO PARTIES

Talk to the child's parents about the special diet before the party. You can suggest foods that your child will eat if some of the foods to be provided do not fit in. Most parents will be happy to make changes if they have enough notice and know exactly what to do. Have lists of suitable snacks, biscuits or lollies as well as cake and icing recipes at hand if asked for these. If the party is at a fast food outlet or at an outside venue suggest appropriate foods, such as hot chips or provide favourite foods for your child to snack on.

Explain that your child should not eat the contents of the lolly bag, any chocolate given as prizes, or the cake (unless it is milk free). You are welcome to provide suitable alternatives so your child can still take part.

Give your child a substantial snack or early meal before the party. A full tummy will take the emphasis off food and make it easier for your child to not eat the unsuitable foods available.

### EATING AWAY FROM HOME

Plan ahead! Whenever possible be prepared – always carry a suitable packaged snack with you just in case. Some quick and easy snacks include fruit, biscuits, cereal pieces, sandwiches and soy drinks.

When eating out for meals it can be useful to call or email ahead and find out about the choices available. Some fast food outlets have information about their products available on their website.

## 14. Getting enough calcium

Calcium is required in everyone's diet for the normal development, structure and strength of teeth and bones and for the proper functioning of the neuromuscular and cardiac systems. While an adequate intake of calcium is important throughout life, the early teenage years appear to be an ideal time to maximise bone health. There appears to be an increased risk of fracture in teenage children who have low calcium intakes.

In the longer term, inadequate calcium is associated with low bone density. This can lead to osteoporosis which is quite common in older men and women in Australia and New Zealand and increases the risk of bone fracture.

People with galactosaemia are at risk of low bone density because of the life-long milk free diet. Reduced oestrogen levels in women with galactosaemia can also diminish bone strength.

The best way to maintain bone health is to:

- Ensure intake of calcium is adequate from diet or supplements.
- Be active with regular weight bearing activities like team sports and walking.
- Discuss hormone levels with your doctor (teenage girls and women with galactosaemia).

If prescribed a calcium supplement, take it as directed by your doctor, nurse practitioner, dietitian, or pharmacist.

### Calcium rich foods

Calcium is found predominantly in milk and milk-based foods so people with galactosaemia need other non-milk sources of calcium.

SOURCES OF CALCIUM INCLUDE:

- Soy drinks fortified with calcium – aim for ~120mg of calcium per 100ml. Alternatively, try calcium fortified rice drink – however be aware that it is lower in calories and protein.
- Soy products such as desserts, yoghurts, ice confection and soy cheese – ensure calcium fortified.
- Canned fish – make sure the bones are included. Salmon and sardines are good sources.
- Tofu, particularly if calcium coagulated – check the label.
- Green vegetables – particularly the Chinese cabbages bok choy and flowering cabbage.
- Permitted matured cheeses.
- Sesame seeds that have not been dehulled and tahini made from these seeds.
- Breakfast cereals with added calcium – check the ingredients list for sources of galactose.

TIPS TO ENCOURAGE YOUR CHILD TO TAKE SOY DRINKS AND PRODUCTS:

- Offer soy drinks and water only as regular drinks. Limit fruit juice and avoid cordial and soft drink.
- Try flavoured soy drinks (make sure they are fortified with calcium).
- Use soy yoghurt, soy custard and soy ice cream, as snacks and desserts.
- Encourage other family members to have soy drinks and products.
- Use soy drinks in cooking and in the preparation of foods.
- Make home-made milkshakes, thick shakes and smoothies using calcium fortified soy products.

## Calcium supplements

Calcium supplements come as

- Tablets which can be crushed, swallowed, chewed or in some cases dissolved in water or juice.
- Soft chewable capsules
- Powders

It is usually preferable to use a supplement that also contains vitamin D. They are available over the counter in pharmacies and supermarkets or sometimes on prescription.

Ask your dietitian for an up-to-date list of calcium supplements.

## Checking calcium intake

You can check dietary calcium intake by copying this page and completing the table. It will be more accurate if you complete the record over several days and take an average. Compare the total you get with the recommended amounts.

DAILY CALCIUM INTAKE:

Food	Calcium content	Amount consumed	Calcium (mg)
Soy drink with added calcium	300mg / 250ml		
Soy yoghurt and custard	300mg / 200g		
Matured cheese	300mg / 40g (40g = 2 cheese slices)		
Tofu	450mg / 125g if calcium-set 170mg / 125g if set with magnesium or nigari		
Sardines	285mg / 5 sardines		
Salmon	280mg / ½ cup		
Tahini paste	69mg / 1 tbsp		
Green Vegetables e.g. bok choy, Chinese flowering cabbage, spinach	70mg / 100g raw		
Calcium supplement	Check label		
<i>Other</i>			
		Total	

## HOW MUCH CALCIUM?

Aim to achieve the following intakes each day. If intake is less than recommended, discuss this with your dietitian and doctor. It is important for everyone in the family to get enough calcium in their diet.

Age	Recommended Daily Intake (FAO:WHO 2001)
1-3	500mg
4-8	700mg
9-11	1000mg
12-18	1300mg
19-70 (men)	1000mg
19-50 (women)	1000mg
Women 51 and older, and men older than 70	1300mg

## 15. Galactosaemia information sheet for caregivers and schools

Galactosaemia is an inherited condition in which the body is unable to metabolise a sugar called galactose. The treatment involves a lifelong strict avoidance of galactose - mainly found as a part of lactose (the sugar in milk). Unlike some diets for lactose or milk intolerance, small amounts of foods containing lactose are not allowed in the diet. Except for the diet, a child with galactosaemia is like any other child. Speech delay can sometimes be a consequence of the disorder, so please discuss any concerns you have with the child's parents or carers.

### About the diet for galactosaemia

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The diet for galactosaemia avoids milk and foods that contain milk or lactose. This requires checking food labels for lactose containing ingredients.

The child's parents are very familiar with the diet and may provide:

- A list of foods that their child can eat.
- A supply of biscuits, cakes or lollies that can be substituted for birthday cakes.
- Alternatives for cooking classes or theme days.

Please speak to the child's parents or carers about any occasions when suitable foods may need to be provided.

### Young children

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Young children need supervision to ensure that they do not eat the wrong foods. Socialising in day-care, pre-school and kindergarten is important for children to learn that all people are different. Children who have been brought up on a special diet are usually very responsible about the foods they can and cannot have. They need to feel confident about refusing food that is not suitable for their diet.

### What will happen if the child eats the wrong food?

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In many cases, no symptoms will occur. We do not yet know what level of galactose intake causes problems. It is likely that a small amount of galactose containing food eaten by mistake will not cause problems, as long as the child is usually strict with the diet. However, if children with galactosaemia consume lactose regularly this could affect their long term health. It is not an emergency if the child eats the wrong food, but the parents or carers should be informed, and steps taken to avoid it happening again.

### Sample letter for parents of a friend of your child

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Dear .....

..... is a classmate of your child. She/he has a rare metabolic condition called galactosaemia, which means her/his body cannot break down galactose from food in the usual way. The treatment is lifelong avoidance of galactose which is mainly found in lactose in dairy products. The condition is not the same as milk allergy or lactose intolerance.

The treatment is avoidance of all foods containing milk. We are happy to send snacks and food if you would like her/him to come and play or to a party. We are happy to talk to you about what food to give her/him if you would like to.

Kind regards,

Phone:

## 16. Glossary of terms

### BLOOD AND URINE TESTS

Laboratory tests can measure galactose and galactose-1-phosphate levels in the blood. Galactitol, an abnormal breakdown product of galactose, is also sometimes measured in the urine. If galactose is eaten by mistake then these levels go up. Even on a very strict diet some children have higher galactose-1-phosphate levels than others.

### CALCIUM

A mineral which is important for strong bones and teeth. The major source of calcium in the Australian and New Zealand diet is milk and milk products. Children and adults on lactose or milk-free diets need to be careful they have an adequate calcium intake from other sources. Calcium supplementation is often recommended.

### ENERGY

The capacity for the body to do work. The body derives its energy from the carbohydrate, fat and protein in food. A kilojoule or calorie is a measure of energy.

### ENZYME

A chemical compound which is involved in the change within the body of one substance to another. For example the enzyme galactose-1-phosphate uridyl transferase helps change galactose to glucose in the body. Another example is the enzyme lactase that splits lactose into glucose and galactose.

### GALACTOSE

A single sugar found in foods and which is also made in the body. Galactose makes up half of the double sugar lactose, which is the major food source. It is found, in much smaller amounts, in other foods. Galactose is incorporated into the structure of the brain and some other organs. In galactosaemia, excess galactose cannot be changed into glucose.

### GENE

A unit of inheritance, which occur in all cells in the body and forms the “blue prints” for all the body processes and traits, such as blood group, hair colour etc.

### GENETICS

The branch of biology dealing with the process by which traits are passed from parents to children.

### INFANT FORMULA AND MILK SUBSTITUTES

These are an important source of nutrients for babies and children with galactosaemia who cannot have breast milk, standard infant formula or cow’s milk. The selected product must be low in both galactose and lactose and appropriate for the age of the child.

### INHERITANCE

The transfer of traits and characteristics from parents and ancestors to children.



## METABOLIC TEAM

Metabolic teams vary depending on where you live. Team members may include a Doctor, Dietitian, Nurse Practitioner, Nurse, Social Worker, Newborn Screening Biochemist, Laboratory Scientist, Psychologist. The team advises on the care of galactosaemia in outpatient clinics or on admission to hospital. Your child may also see a Speech Pathologist, Endocrinologist and/or Gynaecologist.

## METABOLISM

The chemical and physical reactions that occur in the body to support growth, maintenance and other bodily functions.

## NEWBORN SCREENING (previously called the Guthrie Test)

A blood test done on all newborn babies in Australia and New Zealand, which is used to diagnose galactosaemia (and some other rare disorders). Not all states in Australia screen for galactosaemia.

## NUTRIENTS

Components of food that are used as an energy source or other chemicals used by the body that are important for health.

## RECESSIVE

Genes usually occur in pairs. A gene which is altered (by "mistake" or mutation), but which does not affect the carrier in the presence of a normal gene is said to be recessive. In recessive disorders, such as galactosaemia, a person is only affected if both copies of a pair of genes carry a mutation.

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