

Australasian Society for Inborn Errors of Metabolism

# Handbook for Galactosaemia

Australasian Society for Inborn Errors of Metabolism.  
Galactosaemia Handbook

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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PKU Handbook. Australasian Society for Inborn Errors of Metabolism 2005

Low Protein Handbook. Australasian Society for Inborn Errors of Metabolism 2007

Bottle Feeding – A guide to safe preparation and feeding of infant formula. Centre for Health Promotion & Department of Nutrition; Children, Youth and Women's Health Service, Adelaide, 2008

'Learning to Talk' Parent Easy Guide No 33 Parenting SA, Centre for Health Promotion; Children, Youth and Women's Health Service, Adelaide, 2010

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## Introduction to Second Edition 2010

This Handbook replaces the first ASIEM Handbook for Galactosaemia published in 1998. Since that time research has supported the degree of dietary restriction proposed by the Handbook which allowed a wider range of foods than previously allowed in Australia and New Zealand, but highlighted some unusual sources. Since that time there have 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.

Changing from lactose rich breast milk or formula to lactose free formula is very successful in treating the severe effects of galactosaemia in the newborn period. However this, and the galactose restricted diet, however strictly followed, does not prevent the later emergence of long term complications in some children with galactosaemia. The cause of these problems is still not fully understood. Ongoing research will hopefully provide more answers and may lead to future changes in the treatment of galactosaemia. For now a galactose restricted diet is recommended for life. In managing this diet it is important that there is not just a focus on restriction but that nutrient needs, particularly for calcium, are met and food is enjoyed.

### DISCLAIMER

These information sheets are only intended to provide readers and their families with a guide to galactosaemia. They are not intended to substitute for medical and nutritional advice from clinicians and qualified dietitians, preferably those with an expertise in galactosaemia. It is not intended that any person should prefer to rely on the information contained in this book, rather than choose to consult a clinician or dietitian. Whilst every effort has been made to ensure the accuracy of these information sheets, the responsibility for their use resides with the user and not the Australasian Society for Inborn Errors of Metabolism, Editors, Contributors or Members of the working party.



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## 1.1 ABOUT GALACTOSAEMIA

### WHAT IS GALACTOSAEMIA?

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 all animal milks. (Including breast milk, most infant formulas, cow's milk, goat's and sheep's milk.)

There are several types of galactosaemia with the most common being known as classical galactosaemia. Galactokinase deficiency and galactose epimerase deficiency affect different parts of the breakdown pathway of galactose and have different symptoms.

Duarte galactosaemia is a very mild form of galactosaemia which does not require or benefit from treatment.

Your child has .....

### WHAT EFFECTS DOES GALACTOSAEMIA HAVE?

Newborn babies with the most severe form of galactosaemia may be very sick as a result of the disease. Some of them have severe jaundice (yellow colouring/pigmentation or discolouration of the skin) and others can have serious infections. Even if they don't become very ill, many babies with galactosaemia remain jaundiced longer than other babies, and may initially have difficulty gaining weight. Once they are fed with a formula without galactose or lactose, these problems improve rapidly. Some babies with galactosaemia have small cataracts but these disappear quickly once treatment is started.

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

- About half the children with galactosaemia have learning difficulties particularly with speech. This seems to be because they have difficulty in processing information. Speech therapy can be very helpful. Some children will have more severe speech and learning problems and may benefit from appropriate learning support.

- Girls can have specific problems because of the effect of galactose (or one of its breakdown products) on the ovaries, affecting hormone production and fertility (sometimes called ovarian failure). Despite this some women with galactosaemia have successfully had babies of their own.

- Children with galactokinase deficiency are at risk of cataracts. Those with galactose epimerase deficiency (which is very rare) can have similar symptoms to classical galactosaemia.

## HOW IS GALACTOSAEMIA TREATED?

### A Galactose Restricted Diet

At present there is no cure for galactosaemia. The treatment of galactosaemia is currently based on a diet excluding foods that are the major sources of galactose. This is very successful in treating the severe illness 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. At the current state of knowledge it is recommended that the diet for galactosaemia is for life.

The major food source of galactose is from lactose. Lactose is a disaccharide (double sugar) present in human and animal milks. That is why babies with galactosaemia must not be breastfed or fed standard infant formula and a special formula is needed.

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 the 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. Recommendations about avoidance of foods containing trace amounts of galactose varies around the world. In Australia and New Zealand we recommend only avoiding chick peas, large amounts of other legumes, fermented soy bean products, and offal. This level of diet restriction has been used since 1998 throughout Australia and New Zealand.

Children with galactosaemia using these recommendations have not been found to have elevated levels of galactose metabolites in their blood since changing from a more restricted 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.

## 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.

## HOW IS GALACTOSAEMIA DIAGNOSED?

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 uridyl transferase 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.

## HOW DID MY CHILD GET GALACTOSAEMIA?

Galactosaemia is an inherited condition caused by faulty genes. It is uncommon. Around 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. People with galactosaemia are born with a deficiency of one of the enzymes needed to process galactose within the body.

## WHAT ABOUT ACCIDENTAL EXPOSURES TO GALACTOSE?

Although the diet aims to be very low in galactose and is needed for life, mistakes do happen by accident or misunderstanding. There is unlikely to be any immediate effect of this, except in the young baby. If your child accidentally has small amounts of lactose, the long-term effect is thought to be minimal.

## LACTOSE FREE MEDICATIONS

Medications, particularly tablets, often contain lactose. It is important that any medications used, where possible, are lactose free to prevent reduce accidental exposure to lactose. Your pharmacist can help you with this.



## 1.2 WHAT DOES THE DIAGNOSIS OF GALACTOSAEMIA MEAN FOR MY BABY AND OUR FAMILY?

### Your feelings as parents around the diagnosis

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.

The first concern a parent has for their newborn baby is his or her health and well being. 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.

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

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 is rare and has occurred without knowing. It can help to remind yourself that the diagnosis is not something you could have avoided, nor is it the result of anything you have done.

## About Feeding Your Baby – Important Information for Breast Feeding Mothers

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 (the local contact number is in the phone directory)
- La Leche League

It is likely that you will have symptoms of engorgement, as your breasts adjust to not producing milk. Remember to also check for lumps in your breasts, soreness, hot spots or redness. Any symptoms of discomfort, pain, tenderness or fever should be reported to your health professional.

- Massage the breast and express a little milk to relieve the engorgement. If you are having trouble doing this express under a warm shower or after applying a warm heat pack or compress.
- If you have any lumps, apply warmth to the area with a warm heat pack, compress or under a warm shower and gently but firmly massage the lump towards the nipple.
- See your doctor if you feel unwell or have a temperature.
- Use cold packs, washed cold cabbage leaves and pain relief in between expressing.
- Wear a firm, supportive, non-nursing bra to help ease discomfort.
- You do not need to restrict fluids to reduce your milk supply but having less salt in your food may help to relieve the fullness.
- Continue to cuddle and comfort your baby, giving attention and re-assurance. Some mothers find it helpful to have other family members formula feed the baby initially.
- For some mothers, milk may continue to be present in the breast for several weeks.
- "Dry up" medications are not recommended because of increased reports of serious side effects.

Once the practical difficulties of ceasing breastfeeding are over and you see that your baby is content on the formula, take time to acknowledge feelings of loss that you have. Discuss these with a supportive family member, friend or health professional. Develop strategies and answers to deal with situations and comments and remember that there is no other alternative for the health of your baby.

### One step at a time

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

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 other families who have children with galactosaemia if this is what you would like.

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.

## Explaining Galactosaemia

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.

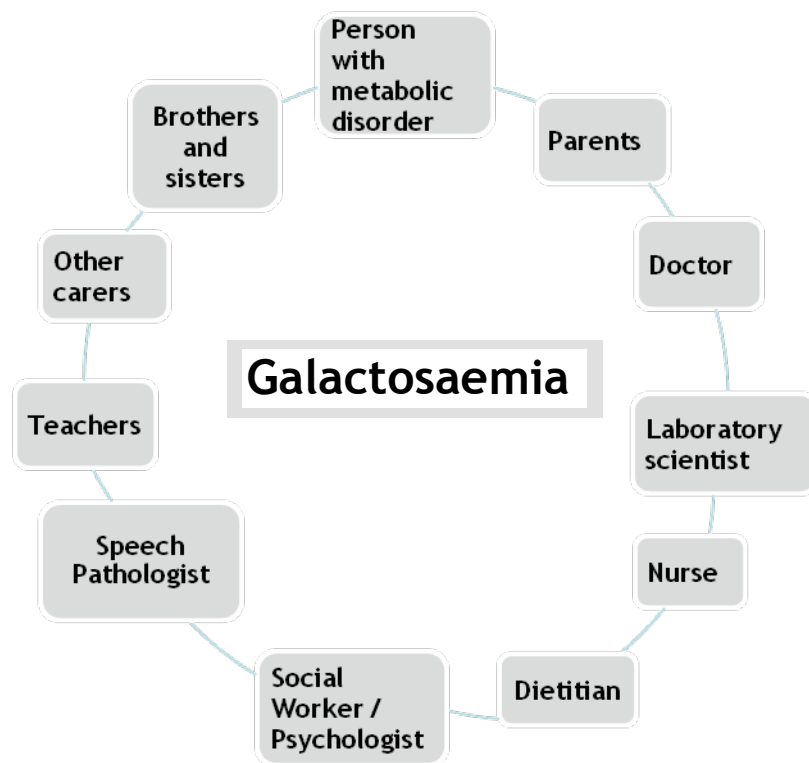
You may feel ready to tell family and friends about the diagnosis soon after it is made. But you may want to avoid telling too many people until you have adjusted to the diagnosis and the extra responsibility it brings. This allows you to let others know about the condition in your own time. Telling 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.

As your child develops it will be important to talk about the diet, any special needs and what galactosaemia is with your child

## A team approach

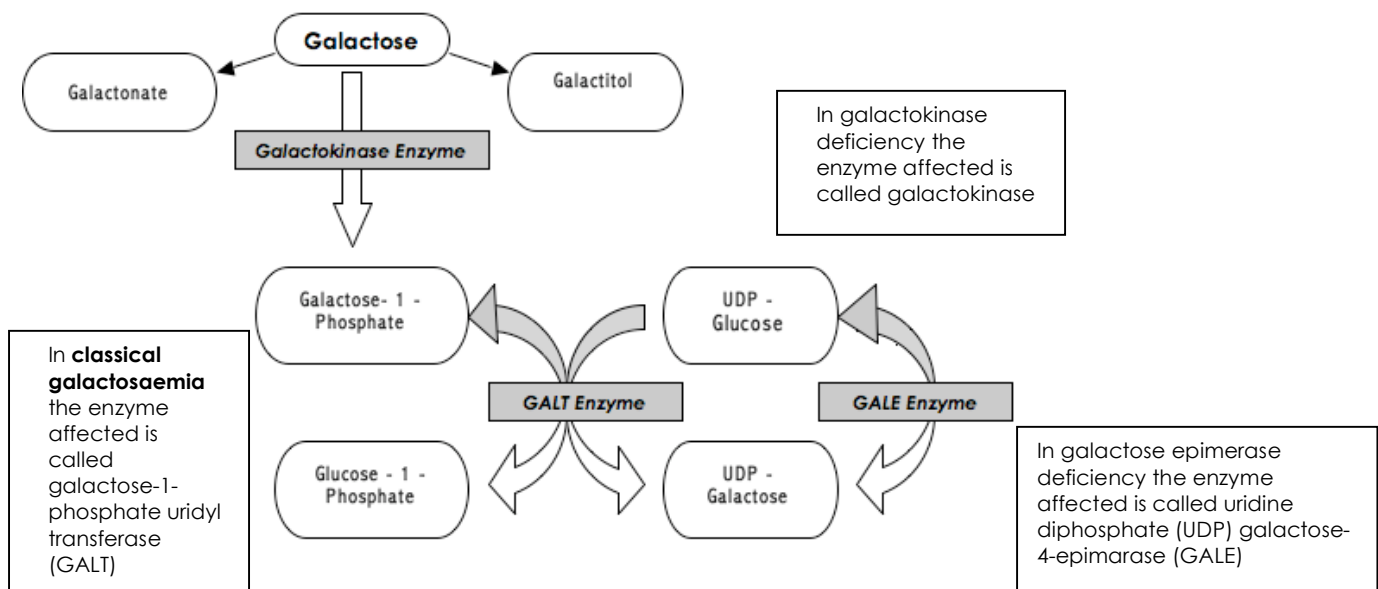
Galactosaemia is not something parents can manage on their own. Even though it can sometimes be difficult to accept guidance on something that seems basic, such as feeding your baby or child, it is important to follow the instructions and recommendations of the metabolic team. It can also be difficult to accept that your child needs specialist help such as speech therapy but early intervention can improve outcomes.

**The metabolic support circle: you will not be alone in managing your child's disorder:**



## 1.3 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.



- Research is ongoing to find improve the management of galactosaemia.
- Blood levels of galactose-1-phosphate (Gal-1-P) have limited use as although they may detect major changes in diet, low levels do not necessarily result in a good outcome. This is because in galactosaemia levels of galactose, galactose-1-phosphate, galactitol and galactonate all increase. It is not understood which, if any, are important for the problems caused by galactosaemia. Increased levels of galactose-1-phosphate may affect other important reactions within the body.
- 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' and research suggests that adults produce around 1000 – 2100 mg galactose each day.
- Overly strict compliance with diet does not seem to improve outcomes such as speech and learning difficulties in galactosaemia.

## 2. GENETICS AND GALACTOSAEMIA—FREQUENTLY ASKED QUESTIONS

### Will other family members be affected?

Other family members can be tested for the disorder and any further babies 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 during a further pregnancy (prenatal 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. Discuss these issues with your metabolic doctor or a genetic counsellor.

### What is the role of genes?

Galactosaemia is caused by a faulty gene. 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 a faulty gene from each parent.

### What happens when genes are faulty?

Everyone carries some faulty genes, usually without knowing which ones. A person who has one faulty gene, but is not affected by it, is called a 'carrier' or 'genetic carrier'. Problems will arise when two people with the same faulty gene have children, as their child may be affected.

### How do faulty genes cause galactosaemia?

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?

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 faulty gene, each egg or sperm cell will carry either a faulty or a regular gene. If, by chance, both parents carry one copy of the same faulty gene, any baby conceived has:

- a one-in-four chance of inheriting two copies of the faulty gene
- a two-in-four chance of becoming a healthy carrier like their parents
- a one-in-four chance of not carrying the faulty gene at all

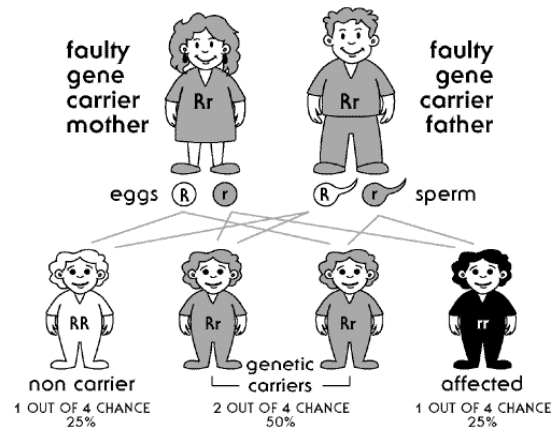
The diagram on the next page describes the inheritance of galactosaemia.

### 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 is difficult and not always accurate. Prenatal testing is usually possible. Discuss testing with your metabolic doctor or a genetic counsellor.

## Inheritance of Galactosaemia



In autosomal recessive inheritance both parents are unaffected genetic carriers for the condition. The faulty copy of the gene containing a recessive mutation is represented by 'r'; the correct copy of the gene by 'R'. If both parents carry one faulty galactosaemia gene 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 8. Available from <http://www.genetics.edu.au> (Accessed June 2010)*

### 3. FEEDING YOUR BABY WITH GALACTOSAEMIA

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

The formula recommended for your baby with galactosaemia will give your baby all of the nutrients he or she needs during the first 4 to 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.

If you were breast feeding when galactosaemia was diagnosed seek help on comfortably reducing your breast milk supply and coping with your emotions at this time. See chapter 1.2, page 10.

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 4 and 6 of this handbook.

#### CHOOSING AN INFANT FORMULA

The following infant formula are suitable for your baby from birth:

Soy based infant formula

S26 Soy	(Wyeth)
Isomil	(Abbott Australia)
Karicare Soya Infant Formula	(Nutricia)

There are no 'Follow On' soy formula available in Australia and New Zealand, but all of the formulas listed are suitable to use after 6 months of age.

Specialised Formula

Sometimes your doctor may prescribe a specialised formula. The following are galactose free:

Neocate	(Nutricia)
Elecare	(Abbott Australia)

The specialised formulas Alfare (Nestle) and Pepti Junior (Nutricia) are based on cow's milk and may contain trace amounts of galactose.

Lactose free ("LF") formula

Lactose free formulas (Karicare Delact and S26 LF) may also contain trace amounts of galactose. These are **not** recommended for managing galactosaemia.

Standard baby formula, "HA" formula and goats milk formula all contain galactose and are not suitable for a baby with galactosaemia

## MAKING UP INFANT FORMULA FOR YOUR BABY

Follow the mixing instructions on the formula tin accurately. It is important that you only use the scoop that came with the tin of powdered formula and add the right number of level scoops to the correct amount of water.

Make up a little more formula than your baby usually drinks to allow for times when your baby is hungrier than usual.

Make sure that bottles and equipment used to make up feeds are sterilised, and that you use cool, boiled water to make the feeds.

Wipe down the bench where you are going to make the formula with a clean cloth.

Always wash and dry your hands thoroughly before handling the bottles and equipment, making up feeds or feeding your baby, as well as after every nappy change.

### How much formula should I make up?

The safest way is to make up one bottle at a time. The table on the can of formula gives you a guide for your baby's age and how much he or she might drink. Do not worry if your baby takes a little more or less than this, as each baby is different. Your baby might even take different amounts of formula at each feed.

If you have to make up more than one feed, boil the water you are going to use (for all babies up to 12 months). Then pour out the cool, boiled water into separate clean and sterilised bottles. Put them in the fridge, and take them out one at a time. When you need them, warm the water first. Then add the powder. Shake the bottle well.

If you have to make up extra bottles of formula in advance, put them in the back of the fridge (where it is the coldest), not the door.

#### **Throw out:**

- **Any formula left in the bottle after a feed.**
- **Any unused water or made up formula after 24 hours.**

### How much formula should I offer my baby?

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.

As a guide, babies usually drink between 125 and 170 ml of formula for each kg of body weight each day. This decreases once solid foods are introduced. Get your baby weighed regularly. If your baby is gaining weight as expected and has 6-8 wet nappies a day he/she is getting enough formula.

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.



## Warming up your baby's formula

Many babies will drink feeds straight from the refrigerator or you can warm a bottle by standing it in a container of warm (not boiling) water for a few minutes. After warming a feed and before using it, check the temperature of the formula by shaking the bottle and then sprinkling a little formula onto your wrist. No heat should be felt. This is very important if you choose to warm feeds in a microwave oven, as there can be hot patches of formula. These can cause burns to your baby's mouth and throat. This is why heating bottles in a microwave oven is discouraged.

If you are going out, take boiled water and formula powder with you to make up feeds when needed. A wide necked vacuum flask of warm water can be used to warm feeds.

If you need to carry ready made formula, take the refrigerated feeds in a cool box or polystyrene container with an ice pack to keep them cool and use the formula within four hours.

## More Information?

More information about making up baby formula may be found on  
<http://www.cyh.com/library/bottlefeeding.pdf>

New Zealand has specific guidelines regarding use of tank and bore water, and these may be accessed on the following website: [http://www.moh.govt.nz/moh.nsf/pagesmh/7756/\\$File/0-2-food-and-nutrition-guidelines-may08.pdf](http://www.moh.govt.nz/moh.nsf/pagesmh/7756/$File/0-2-food-and-nutrition-guidelines-may08.pdf)

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.

## 4.1 DIET FOR GALACTOSAEMIA

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.

Lactose =  
Glucose +  
Galactose

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.

Your family's diet will be healthier, and 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:

Current Australian and New Zealand recommendations allow foods containing minimal amounts of galactose in the diet, whilst avoiding the following foods or ingredients:

All animal milks - including human breast milk, cow's milk, goat's, sheep, buffalo 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.

Chick peas.

Intake of other legumes, fermented soy bean products and offal meats such as liver and kidney should be limited as these contain some galactose.

### LACTOSE IN FOODS

Lactose is present in human and all animal milks, and in most products made from them. Many manufactured products contain added lactose, milk or milk products. Lactose free and lactose reduced food usually still contains galactose. It is important to always carefully check ingredient labels on packets and cartons.

### LACTOSE REDUCED AND LACTOSE FREE MILKS AND YOGHURTS

Lactose reduced and lactose free milk, cream, yoghurt and custard are **not** suitable for galactosaemia. This is because they still contain galactose.

## 4.2 LABEL READING

Food manufacturers have to include a lot of information on their food labels. The labels can be complicated to read and understand but 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.

When you buy a packaged food you should carefully read the ingredient list on the food packet to check if the food contains sources of galactose. There are many different names that milk and lactose can be called in the ingredient list, some of which are simple to identify, and others that are not so obvious.

Food ingredients that are sources of galactose – avoid these in Galactosaemia.

Ingredients from milk:	
butter, butter fat	lactose
buttermilk	margarine (containing whey or milk products)
casein, calcium caseinate, sodium caseinate	milk, non fat milk, skim milk milk protein
cheese*, cheese powder	milk solids, non fat milk solids, skim milk solids
cream, sour cream	milk sugar, sugar of milk
fruche, fromage frais	skim milk powder
ghee	whey solids, whey syrup sweetener, hydrolysed whey sugar, whey powder
hydrolysed protein (if derived from milk protein)	yoghurt
ice cream	
Other ingredients:	
fermented soy products eg miso, tempeh, natto	
chick peas, chick pea flour (besan)	
offal eg liver, kidney, brains, pate.	

\* Some matured cheese may be allowed in the diet. During the maturing process, galactose is broken down so that only trace amounts are left. In manufactured food products it is wise to avoid those that contain cheese on the ingredient list because this may not be a matured cheese and may contain significant amounts of lactose.

NB: If a food does not have a label (eg 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 are able to 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.

### FOOD INGREDIENTS THAT ARE NOT SOURCES OF LACTOSE OR GALACTOSE

The following ingredients are not a source of lactose or galactose and can be included in the diet:

cocoa butter	coconut milk
lactic acid (additive number 270)	lactate (additive number 326, 327)
peanut butter	non-dairy cream
caramel (although caramel may be used in products that also contain milk – check the label)	

## WHAT HAS TO BE ON A FOOD LABEL?

All food labels have to 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 a number of 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 in-going 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.

## 4.3 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.

***Just in case*** – keep a supply of a lactose-free medicine for fever or pain (eg paracetamol) at home.

## 4.4 GALACTOSAEMIA DIET CHECK LIST

MILK, MILK PRODUCTS AND SUBSTITUTES		
ALLOWED	CHECK INGREDIENTS	NOT ALLOWED
<p>Soy Based Infant Formula:</p> <p>S26 Soy Isomil Karicare Soya Infant Formula</p> <p>Other Specialised Formula:</p> <p>Neocate &amp; Elecare</p>		<p>Breast milk Cow's milk based infant formula 'HA' infant formula Goat's milk based infant formula</p> <p>Other infant formula not prescribed by Metabolic Team</p> <p>Alfare, Pepti Junior, Karicare Delact and S26 LF are based on cow's milk protein and may contain trace amounts of lactose</p>
<p>Soy drinks (with added calcium): eg So Good Regular</p> <p>These may be introduced in the preparation of food from 6 months of age and as a drink from around 1 year of age</p>	<p>Soy drinks (should contain approximately 120mg calcium per 100ml)</p> <p>Choose soy drinks containing soy protein isolate as the protein source</p> <p>Do not use reduced fat varieties for children under 2 years of age</p>	<p>Cow's milk, goat's milk, sheep's milk (fresh, UHT, powdered, evaporated, condensed)</p> <p>Reduced lactose milk eg Digestelact, Lacto-Lo, Zymil, Harvey Fresh</p> <p>Lactose free cream Lactose free yoghurt</p> <p>Fermented milk-based drinks</p> <p>Home made soy milk</p>
<p>Rice and Oat Beverages</p> <p><b>NOTE</b> - These are not nutritionally adequate for young children and should only be used under medical supervision</p>	<p>Avoid rice beverages containing chick pea protein</p> <p>Check rice or oat beverages contain approximately 120mg calcium per 100ml</p>	<p>Home made cereal and nut milks</p>

CHEESE		
ALLOWED	CHECK INGREDIENTS	NOT ALLOWED
<p>The harder and older the cheese the less galactose it will contain: best choice is hard mature cheddar or imported gruyere, emmental and jarlsberg</p> <p>Small amounts of hard blue vein cheese, gouda, edam, parmesan, romano, pecorino, camembert, brie and Swiss cheese may be acceptable. See Ch 4.5, p28.</p> <p><b>Discuss introduction of cheese with your dietitian.</b></p>	<p>Soy cheese</p> <p>Note some soy cheese contains whey or casein which may contain some lactose</p>	<p>Cream cheese, cottage cheese, processed cheese, cheese spread, ricotta, haloumi, feta, neufchatal are all likely to contain significant amounts of lactose</p> <p>Goat's milk, goat's cheese, sheep's milk, sheep cheese, buffalo milk and buffalo cheese are all sources of lactose</p>

DESSERTS		
ALLOWED	CHECK INGREDIENTS	NOT ALLOWED
Desserts made from milk substitutes and other allowed ingredients  Jelly	Soy or tofu based ice confections, yoghurt and desserts  Soy yoghurt does not usually contain lactose. However because it is a fermented product it may contain some galactose released from fermentation of the naturally occurring sugars in the soy. <b>Discuss with your dietitian.</b>	Cream  Custard made from milk  Dairy desserts Fromage frais (eg Fruche) Ice cream, ice confection  Yoghurt (made from cow's milk, goat's milk, sheep's milk)  Lactose free yoghurts and other lactose free desserts made with cow's milk

BREADS, CEREALS, PASTA, RICE, CAKES and BISCUITS		
ALLOWED	CHECK INGREDIENTS	NOT ALLOWED
Flour made from wheat, rye, barley, rice, maize (corn), soy, arrowroot	Bread (all types), muffins, crumpets, pizza base	Any bread, cereals, pasta, rice, cakes and biscuits, crackers, rusks containing milk or lactose  Besan flour (made from chick peas)
Bran, cornmeal, wheatgerm, oats, rolled oats, ground rice, semolina, tapioca, sago	Breakfast cereal, baby cereals Custard powder, dessert mixes	
Pasta, noodles (plain), cous cous, rice, barley, cracked wheat, polenta	Pasta, noodles or rice meals (packet, frozen or canned with flavourings)	
Homemade biscuits and cakes from allowed ingredients	Biscuits, crackers, rusks, cakes, scones, pastries and slices (icing may also contain galactose)	

SNACKS		
ALLOWED	CHECK	NOT ALLOWED
Plain boiled sweets Milk-free chocolate Milk-free carob Nuts in shell Home-made popcorn	Chocolate, carob, lollies, caramels  Flavoured or salted nuts, popcorn or snack foods (lactose may be used as a carrier for salt or spices)  Flavoured potato crisps and corn chips  Muesli and cereal bars	Chocolate and lollies containing milk or lactose  Any snacks containing sources of milk or lactose

FRUITS, VEGETABLES and LEGUMES		
ALLOWED	CHECK INGREDIENTS	NOT ALLOWED
<b>Fruit:</b> fresh, dried, frozen  Fruit juice, fruit juice drinks	Fruit based desserts or pastries  Fruit juice combinations from juice bars	Any fruit based desserts or pastries containing sources of galactose eg apple custard  Fruit smoothies
<b>Vegetables:</b> fresh, frozen, dried or canned in brine or water with no other ingredients	Prepurchased or frozen vegetable or salad dishes  Packet and canned soup mixes	Instant mashed potato  Vegetables served with galactose containing ingredients, cheese sauce  Hot chips and wedges with chicken salt
<b>Legumes:</b> (dried peas, beans, lentils) include in moderate amounts only  See chapter 4.5 "Controversial Foods"	Vegetarian foods or salads  Baked beans	Chick peas, besan flour, chick pea dahl, hummous, felfafal made with chick peas  Fermented soy products, miso, tempeh, natto

MEAT, FISH, EGGS, POULTRY		
ALLOWED	CHECK	NOT ALLOWED
<b>Meat:</b> plain fresh or frozen  <b>Poultry:</b> plain fresh or frozen  <b>Fish:</b> plain fresh, frozen, canned in oil	Meat or chicken in marinades or sauce Corned meat Crumbed or battered foods, schnitzel, fish fingers, chicken nuggets, battered fish, sausage rolls, meat pies Tinned meat and fish in sauce deli meats, manufactured meats eg salami, mortadella, sausages, hamburgers Meat and fish paste	Any pre-prepared meat, chicken, fish or egg dishes containing sources of galactose  Offal (liver, kidney, brain, sweetbreads and pancreas) contain some galactose and should not be eaten in large amounts eg liver pate  Seasoning mixes containing lactose
<b>Eggs:</b> plain or with allowed ingredients	Egg substitute, yolk free egg mix	
<b>REMEMBER:</b> Always check with your butcher, chicken, poultry shop or take away shop about ingredients in sausages and other ready made products.		



THE KITCHEN CUPBOARD		
ALLOWED	CHECK	NOT ALLOWED
Sugar, honey, jam, treacle, golden syrup, molasses, marmalade, maple syrup	Powdered artificial sweeteners	
Vegemite, Promite, Marmite Peanut butter	Meat or fish pastes	Any spreads containing sources of lactose eg Nuttella
Salt, pepper, vinegar, herbs, plain spices, essences  Coconut, coconut milk, coconut cream	Liquid and powdered stock Stock cubes, mixed spices MSG powder Curry pastes Gravy mixes Bottled sauces eg tomato Pickles, chutneys Canned or bottled casserole Pasta sauce and curry bases Canned, bottled or packet soups (as well as checking for lactose also remember to use only moderate amounts of those containing legumes) Salad dressings, mayonnaise	Miso, Tempeh and Natto  Large quantities of soy sauce  For information on soy sauce see ch 4.5 'Controversial Foods'

FATS AND OILS		
ALLOWED	CHECK	NOT ALLOWED
Milk free margarine eg Becel, Nuttalex, Olivani Oil Cophera, lard, dripping	Solid frying compounds  Margarine	Imitation cream Butter Regular margarine Cream Ghee

DRINKS		
ALLOWED	CHECK	NOT ALLOWED
Soy or Rice drink (Plain or flavoured with fruit or cocoa and sugar or other allowed flavouring) Tea, coffee (no milk) Fruit juice, soft drinks, cordials, soda water, tonic water, mineral water	Milk flavourings Ice cream toppings Drinking chocolate	Milk, milk shakes, flavoured milk  Milk flavourings containing lactose, malted milk powder, Milo, Ovaltine

## 4.5 CONTROVERSIAL FOODS

If you read information about the diet for galactosaemia from other information sources such as the internet you will note that some countries restrict many foods that are not restricted in Australia and New Zealand.

Some foods, particularly plant foods (fruits and vegetables) contain small amounts of galactose. In some countries some fruits and vegetables are restricted in the diet for galactosaemia. In Australia and New Zealand current recommendations allow foods containing very small amounts of free galactose or bound galactose 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 the more restricted diet which was previously used. Research in America and also in Holland has also supported the Australian findings that intake of fruits and vegetables makes no difference to galactose-1-phosphate measurements. This is probably because the body is able to 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 and more enjoyable and reduces the risk of inadequate intakes of nutrients during times of rapid growth and development.

### Controversial Foods Containing Small Amounts of Free or Bound Galactose:

The table below compares the amounts of free galactose in many of the controversial foods with foods that are very high in galactose. This explains why we do not restrict fruit and vegetable intake in Australia and New Zealand.

#### The Free Galactose Content of Foods:

FOOD	Galactose mg
<b>Food high or very high in galactose – to be avoided</b>	
Lactose 1 teaspoon (3g)	1500
Cow's milk 100 ml	2350
Human milk 100 ml	3700
Chick pea (garbanzo beans) (cooked) 100g (2/3 cup)	444
Snack foods with lactose (eg salt and vinegar crisps, 50g packet)	850
<b>Foods with moderate amounts of galactose – to include in limited quantities only</b>	
Baby lima beans (cooked) 100g (2/3 cup)	175
Kidney beans (cooked) 100g (2/3 cup)	153
Lentils (cooked) 100g (2/3 cup)	116
Soy Sauce (20g)	35
<b>Foods with only small amounts of galactose – can be included in diet</b>	
Soy beans (cooked) 100g (2/3 cup)	43
Kiwifruit (100g) 4 fruit	27
Tomato (100g) ¾ medium	23 (maximum reported)
3 serves fruits (apple, banana, orange) and 3 serves vegetables (potato, carrot, green beans)	40
Cheddar cheese 20g (1 slice)	2

## Cheese

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 used up 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 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 the less galactose will be present. Hard matured or vintage cheddar is the best choice and is a good source of calcium.

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

The following cheeses vary in galactose content depending on their production method: parmesan, romano, pecorino, gouda, edam, camembert, brie and Swiss cheese. Small amounts may be tolerated - discuss with your dietitian.

## Cocoa

Cocoa may contain bound galactose, but is considered acceptable for people with galactosaemia. But remember that a bar of chocolate usually contains milk.

## Eggs

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

## Fermented foods (eg pickles, sauerkraut)

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 that intake of these fermented foods is acceptable.

## Gums and fibres (eg acacia, agar, carrageenan, carob, guar gum, gum arabic, locust bean gum, tragacanth gum, xanthum gum)

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)

Research has suggested that some legumes contain significant amounts of free galactose as well as galactose in the bound form. It is recommended to avoid chickpeas and any fermented bean products (see soy beans) except for small amounts of soy sauce and use other dried or canned beans, peas and lentils in moderation. Fresh or frozen peas, green beans, snowpeas and snap peas do not contain significant amounts of galactose

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

These will contain some galactose but it is unknown how much they contribute to intake. These are restricted by some clinics and limited intake is recommended. Note that pate is made with liver and intake should be limited.

### Seeds and nuts

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

### Soy beans and soy products

Soy beans are acceptable in the diet along with tofu and soy drinks. It is probably wise to avoid fermented soy products like tempeh, natto and miso. Galactose may be released from bound sources during the manufacturing of soy sauce but small amounts will not contribute significantly to the galactose intake.

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

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.

## 5. RECIPE ADAPTATION

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 says:	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 4.5, page 27)
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:

Tbs =  
Tablespoon  
  
tsp =  
teaspoon

### Custard

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

250ml soy formula or soy drink  
2 Tbs cornflour  
1 Tbs sugar  
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  
1 ¼ cups soy or rice drink  
2 Tbs plain flour

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, 1 cup of grated matured cheese and ½ cup cooked frozen peas and corn.

### Pikelets

2 cups plain flour  
1 1/2 cups soy or rice drink  
3 tsp baking powder  
1 egg  
75g melted milk free margarine  
1/3 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  
¾ cup soy or rice drink  
1 Tbs milk free margarine

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  
2 Tbs honey

$\frac{1}{4}$  cup sugar  
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.

Copy easy recipes for cakes, biscuits or slices for times when people ask for these.

## Cup Cakes

60g milk free margarine  
1 egg  
2 Tbs soy or rice drink

$\frac{1}{4}$  cup sugar  
1 cup self raising flour  
 $\frac{1}{4}$  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  $\frac{3}{4}$  cup icing sugar mixture and 1 Tbs water.

## Banana Cake

125g milk free margarine  
 $\frac{3}{4}$  cup brown sugar  
2 very ripe bananas, mashed  
1 tsp baking soda  
1 Tbs hot water

$1\frac{1}{2}$  cups flour  
 $1\frac{1}{2}$  tsp baking powder  
1 tsp cinnamon  
 $\frac{1}{2}$  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, no need to peel before grating  
 $\frac{1}{2}$  cup sultanas  
1 tsp baking soda

1 Tbsp hot water  
1 tsp mixed spice  
1 tsp cinnamon  
1 cup flour  
 $\frac{1}{2}$  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	3 Tbs cocoa
1½ cups flour	1 tsp vanilla essence
1 tsp baking powder	1 Tbs malt vinegar
½ tsp baking soda	5 Tbsp oil
½ tsp salt	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	½ tsp baking soda
1 tsp baking powder	2 Tbs hot water
1 cup coconut	125g milk free margarine
1 cup brown sugar	1 Tbs golden syrup
1 cup rolled oats	

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.



## 6. STARTING YOUR BABY WITH GALACTOSAEMIA ON SOLID FOODS

Learning to eat solid foods is a new experience for your baby and the following guidelines give practical handy hints. As parents / carers you will need to know the types of food that are suitable for your baby with galactosaemia. Your clinic dietitian and the chapter “Diet for Galactosaemia” will help you with this. There are lots of foods that your baby can enjoy.

### WHEN WILL MY BABY BE READY FOR SOLID FOODS?

Babies are ready to start solid foods when they are able to sit, with support, and hold their head upright and steady. At this time, they may also be starting to show signs of being interested in what other people are eating.

This usually happens at around six months of age. Babies develop at different rates and some may be ready for solid foods from four months onwards, but should not be given solids prior to four months of age.

Learning to eat solid foods generally takes babies several months as they get used to new tastes and textures.

### HOW TO GIVE SOLID FOODS TO YOUR BABY

Choose a quiet time of day when you and your baby are relaxed after a formula feed. In a quiet place, seat your baby in a secure position facing you. Use a small spoon such as a teaspoon or a baby feeding spoon with smooth edges. Do not add solids to your baby's bottle.

Place a small amount of food on the spoon and hold it to your baby's mouth. Press the spoon gently down on your baby's lips. When their mouth opens, place the tip of the spoon just inside. Babies take the food from the spoon in a sucking motion.

At first, babies may push the food out with their tongue. This doesn't mean they don't like it – just that it is a new experience or taste. Your baby will gradually learn to take the food off the spoon and swallow it. Eating skills improve quickly over a week or two.

All babies have their own ways and preferences where food is concerned. Give your baby solids that suit their feeding skills.

Solid food is introduced in three stages:

- Smooth puree and lump free food
- Lumps and chunks that can be chewed
- Self feeding and finger foods

## Introducing solids - three steps

### Step 1: Smooth puree and lump free food

Start with one food at a time and offer only 1-2 teaspoons at first, gradually increasing the amount over several days until your baby is taking as much as he/she wants.

It is best to start with bland, smooth foods, such as a milk free rice cereal, fruit puree (apple, pear, peach, avocado) or vegetable puree (potato, sweet potato/kumara, pumpkin). Remember to check the label of any manufactured products to check they do not contain milk or lactose.

Gradually increase the amount and variety of food, offering one or two new foods a week. When your baby is having about 1/4 to 1/2 a cup, you can offer foods at a second and then third feeding time.

At around 6 months, introduce purees of meat, fish and chicken. Egg is one of the more common foods that babies can be allergic to so introduce small amounts of cooked egg to start with and if tolerated you can increase the amount. For desserts you can make custard with the formula (see recipes).

Your baby will enjoy the natural tastes of foods so there is no need to add any fat, sugar, salt or spice to their food.

### Step 2: Soft lumps and chunks

At around eight to nine months babies start to make chewing movements, whether or not they have teeth. This is the time to introduce thicker and lumpier foods. You can now give food **before** a formula feed.

Also at this stage:

- foods can be mashed, grated, diced and pureed more thickly than before
- you can increase the variety and amount of foods your baby eats – try casseroles of meat and vegetables, soups, pasta, rice, different breakfast cereals, and custards, desserts and white sauce made with soy formula or soy drink (see recipes)
- offer sips of water from a sipper cup
- keep giving formula from a bottle and small amounts from a sipper cup

### Step 3: Self feeding and finger foods

Once babies are managing lumpy foods they start to become interested in feeding themselves. This usually happens at around ten to twelve months of age, though you can start offering finger foods from seven months onwards.

Babies will be eager to feed themselves with fingers and a spoon if given the chance. When feeding your baby from a bowl use two spoons, one for them to practise with and one for you.

Learning to eat is a messy business! Food will be dropped and spilt. Easy-to-clean baby equipment makes life easier.

If babies get used to sitting in a highchair or at a low table when eating, it keeps them focused on eating and also helps to confine the mess. Placing a plastic mat on the floor makes spills easier to clean up.

### Ideal finger foods include:

- hard dry toast made from milk free bread with or without milk free margarine
- peeled soft fruits
- strips or pieces of cooked vegetables
- pasta
- rusks (check these are milk and lactose free)

Always stay in the room with your baby when he/she is eating finger foods or self feeding. Babies can choke on large pieces of hard foods. If it is possible for a hard piece of the food to be broken off, grate, cook or mash the food eg raw carrot, celery or apple.

### Food Preparation

Cleanliness is important when preparing food as well as formula to reduce the risk of food poisoning. Always wash and dry your hands thoroughly before preparing food. Keep your kitchen and utensils clean. Don't leave warm or hot food sitting around on the kitchen bench. If you want to keep food for later, put it in the refrigerator straight away (once the food has stopped steaming). Reheat chilled or frozen food thoroughly before serving.

There are several ways to prepare foods for your baby:

- Use freshly prepared foods each day. Small amounts can be prepared and cooked on the stove or in a microwave.
- Prepare a larger amount and store it in a small covered container for up to 48 hours in the refrigerator.
- Freeze prepared food in ice cube containers covered with plastic wrap, for use later. Once frozen, tip the cubes into a zip lock bag, remove as much air as possible, seal the bag and label with the contents and date. Immediately return to the freezer to store for up to 3 months. Simply thaw and warm a cube or two of selected foods as needed. This means you can have several varieties to mix and match, once your baby is taking a variety of different foods.
- Try adding herbs and spices to make food tastier but remember that your baby will enjoy the natural tastes of foods and there is no need to add salt or sugar.
- Some family meals will be suitable and checking that they fit with the diet for galactosaemia gives you practise in learning the diet.
- Choose commercial baby foods that are milk and lactose free. Your dietitian will explain how to check food labels. See list of ingredients to check for in "Diet for Galactosaemia".

## Changing from formula to soy drinks

- Keep the bottle for the prescribed soy or special formula.
- Drinks such as water and diluted fruit juice can be offered from a cup but your baby will still need between 600 and 800ml formula a day.
- Soy drinks (choose one containing about 120mg calcium per 100ml) can be used if convenient for cereal, custard and cooking from 8 -10 months. Offer it as a drink from a cup from 1 year of age and gradually replace the bottle feeds of formula with this.
- By 18 months of age your baby should be taking most of their soy drink from a cup.

## How to develop good eating habits

The following suggestions may be helpful:

- establish a meal and snack schedule
- offer food at the table or in a high chair
- discourage eating between meals and snack times
- offer formula at meal times and water at other times
- offer vegetables or fruit at most meals and snacks
- be aware that changes in appetite and intake are normal
- continue to offer healthy choices at consistent times, even though your child may sometimes refuse food or become 'picky'
- have your child sit at mealtimes whether or not they are eating
- stay positive about food and formula and avoid power struggles

Talk to your child about their special diet from a young age.

## Mealtime, family time

Your child should be part of the family meal times from an early age. This will encourage good eating habits and help your child to learn about his or her diet.

Sometimes your child will want some of the foods he or she cannot have. You will need to tell him/her about being "special" and the foods he or she can eat. Always have some foods or alternatives that you can offer, such as fruit or a suitable ice cream alternative or milk free biscuit, as a replacement for those he/she can't have.

Avoid the temptation to have a 'cow's milk free house' - children need to learn what they can and cannot eat at home. This helps them cope when they are out and are faced with food that they cannot eat.

## 7. TODDLER AND PRE SCHOOL EATING

Refusing food is a common but frustrating problem during the early childhood years. When children need to follow a special diet it can be even more difficult.

Aim to have three meals with a snack mid morning and mid afternoon. Don't let your child eat constantly throughout the day. Let your child know that it will soon be time to eat.

Understanding why children don't always eat as well as we would like them to makes it easier to avoid the situation where mealtimes turn into an unpleasant experience for the whole family.

Your child will let you know when he or she has eaten enough. This is the time to stop feeding, even though your child may not have finished the meal. Offer realistic serving sizes. For example, a toddler eats between a quarter and a third of an adult meal. Allow your child to ask for more if they are hungry.

### Seven tips for reducing tension at meal times

#### 1. Respect that your child may not be hungry.

After the age of 12 months children don't grow as quickly, which means their appetite won't be as large as it was in the first year.

#### 2. Watch for growing independence.

As toddlers start discovering that they are independent people, they may express their likes and dislikes more strongly.

It may help to allow your child to choose between two foods, or be involved in preparing the food. Many toddlers want to feed themselves rather than be spoon-fed. Give them their own spoon to try. Offering plenty of finger foods encourages independence.

#### 3. Avoid battles over meals.

Parents often become anxious when their children don't eat, and children quickly pick up on this. Some children refuse to eat, knowing it is an effective way to gain attention.

It is never a good idea to force-feed a child. This often leads to fear of mealtimes and further food refusal. Continue to offer new foods over time. It may take many offers before your child will taste the food, and many tastes before they like it. Praise your child for trying new foods.

Remember to stay calm if your child refuses their meal. Avoid offering alternatives. They will eat more at the next meal or snack.

#### 4. Help your child understand their special diet.

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.

## 5. Create a pleasant mealtime environment.

Parents and siblings can be good role models for young children. Eat meals together as a family as often as possible.

Try to keep the mealtime atmosphere relaxed so that this is an enjoyable time for the family. Turn off the television and try to avoid having family arguments at the dinner table.

## 6. Keep regular routines.

Children respond well to having predictable routines. They need to eat regularly to meet the demands of their growing bodies.

Seat your child at the table for meals. Children have short attention spans. Set aside 20–30 minutes for meals, and 10–15 minutes for snacks. Forcing your child to sit for longer may lead to further food refusal.

## 7. Keep a positive attitude to your child's diet.

It is important for your family and friends to have a positive attitude towards your child's diet.

Allow your child to form their own opinions about their diet. Stay calm and try not to be anxious about how well your child eats.

## Feeding toddlers

Remember that:

- most healthy children will not starve themselves
- 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

## 8. LEARNING TO TALK

Children with galactosaemia may suffer from speech problems, particularly dyspraxia - a speech disorder in which a person has trouble saying what her or she wants to say correctly and consistently. This section gives background information about normal speech development and is from Parent Easy Guide No 33, Parenting SA. This Parent Easy Guide uses 'he' and 'she' in turn. Change to suit your child's sex.

Learning to talk is one of the most difficult and important steps that young children take. It helps them to make sense of the world, to ask for what they need and to be able to get on with other people. If you think about how difficult it is for adults to learn a different language you can get some idea of what it is like for an infant to learn to speak from having no language at all. Language and speech development, like other development, takes place at different rates for different children.

### Steps in learning to talk

#### The early months

Long before they can speak, babies are listening to their parents and carers. They begin to make little noises and sounds which come before speech. If parents and carers imitate these, it is as if they are talking to the baby. This is the beginning of your baby learning to talk. By responding to your baby's needs when she cries, you show that you have heard her and that she matters. This is the beginning of communication.

#### 8-12 months

The early little noises turn into babbling, eg "Da-da-da-da" and "Ma-ma-ma-ma". Babies are beginning to learn what some simple words mean even though they cannot say them, eg "Mummy, Bottle, No". There may be one or two single words. Babies wave "bye-bye" when asked, and can make some gestures. They obey simple requests such as "Give me the ball".

#### 12-18 months

There is much babbling in the children's own jargon. The first single words appear, eg "No, Dad, Dog". Children can point to things that they know when they are asked to. They enjoy songs, music and books. Children know their own names and respond to them.

#### 18 months to 2 years

18 month olds can know and use six or more words. Two year olds may have a 100 or more words. Many of the words may be unclear but the parent or carer can tell what is meant.

Two year olds can say their name.

They can ask for simple things that they need, eg "Drink".

Children start to join words together, eg "Daddy home", "All gone".

They copy the last part of sentences.

They try out different speech sounds and make mistakes.

#### 3 to 4 years

Children begin to ask "What?" and "Why?" questions.

Use sentences with three or four words.

They begin to separate the truth from make-believe.

They can talk about 'Yesterday, Now and Tomorrow' and know what they mean.

Their speech should be understandable most of the time.

They are likely to talk to themselves as they do things.

They can learn and join in simple rhymes and songs.

#### 4 to 5 years

Children learn to adjust their language to the situation they are in. For example they talk differently to their parents than they do to their friends.

They ask 'When?' questions.

They can talk about imaginary situations, eg 'I hope . . .'

They still mix truth and make-believe.

They like to tell stories.

They can hold conversations with their friends and parents.

They will be able to say their name, age and address if they have been taught this.

Four year olds enjoy making up words for fun and using toilet words, eg 'poo', 'bum'.

Their speech is clearer but they still may not be using 'th', 'r', 'z', 's', and 'v'.

## What parents can do

- Talk to your baby right from birth and imitate her sounds.
- Name things and talk about what you are doing. Use simple words and sentences at first with an emphasis on key words.
- Have conversations with your child at some time every day.
- Listen with interest when your child is talking to you. Don't interfere or correct your child's speech.
- Answer questions simply and clearly.
- Allow your child time to get out what she wants to say.
- Talk about pictures in books, and name things in the pictures.
- Sing songs and read rhymes with enthusiasm.
- Take your children to the local library and read some stories to them. Then you can borrow or buy the ones that they particularly enjoy.
- Give a younger child a chance to talk without being interrupted by older brothers and sisters.
- If your child is stumbling over words because he is excited suggest that he tell you slowly. Then listen to him carefully.
- Get down to eye level with your child when teaching a new word so he can see your lips and hear the word clearly.
- For children with a severe hearing loss, it is most important that their hearing loss is recognised before six months of age.

## Be concerned if your child:

- does not react to loud noises by the time she is one month
- does not turn her head to a noise or voice by three months. Hearing problems often cause speech difficulties.
- does not start to make single sounds, eg 'ba ba' by eight or nine months
- does not babble or make other sounds when someone talks to her by twelve months
- is not starting to say single words by twelve months.
- does not understand simple instructions by two years
- frequently repeats sounds or part-words, eg 'Wh-wh-where's my ba-ba-ball?'
- lengthens sounds or gets stuck on words, eg 'm-m-m-m' or 'da-a-a-a-ad'
- is embarrassed or worried when speaking.
- If you have any concerns at any stage about your child's speech, talk to your local child health nurse or your doctor. Your child may need to see a speech pathologist (through local Community Health Centres, Hospitals that provide services for children, or Privately).

## Reminders

Language development needs listening and talking.

Use simple language.

Sit or kneel down so you are on your child's level when she is talking to you.

Spend time reading simple stories and rhymes, looking at picture books and singing songs.

Help your child to notice road signs and billboards.

Learning language is important. It should also be fun.



## 9. EATING AWAY FROM HOME

### 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 (Appendix 1). 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 galactose free.

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 he/she recognises as being allowed in his or her diet - like fruit - not sandwiches, biscuits and snack foods.

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. You could offer to bring along one of your child's favourite foods as a contribution. 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 or any chocolate given as prizes, but is to bring them home. If the cake is not milk free, suggest it be brought home. Once at home have alternatives to swap.

Don't send your child to the party hungry. 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. Many young children are too excited to eat at birthday parties.

## 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 fresh fruit, dry biscuits, dry cereal pieces, half a sandwich and tetra packs of flavoured soy drink.

When eating out for meals try to make the best choices that you can. 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.

Avoid obvious sources of milk. Most breads, baked potatoes, hot chips, plain barbecued or grilled meat and chicken (check whether lactose is used on the skin or in seasoning), green salads (without dressing) and fresh fruit are usually suitable.

## 10. HEALTHY BONES – 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, low intakes of calcium are associated with low bone density (loss of bone strength). 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 as a result 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 each day as directed by your doctor, 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. Milk substitutes, such as soy drinks, supplemented with calcium are a good substitute for milk, and calcium is also found in smaller amounts in fish with bones, legumes, some other soy products and breakfast cereals.

#### Sources of calcium include:

- Soy drinks fortified with calcium – use those with approximately 120mg of calcium per 100ml (read the nutrition label). If soy drinks are not liked, try calcium fortified rice drink as a substitute – however be aware that it is lower in calories and protein
- Soy products such as desserts, yoghurts, ice confection and soy cheese – check the label for calcium content and sources of galactose
- 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 to one small glass per day. 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
- Offer soy drinks and products to other family members – if calcium is added, they are nutritionally equivalent to milk products and taste terrific!
- Use soy drinks in cooking and in the preparation of foods
- Make home-made milkshakes, thick shakes and smoothies using soy drinks and soy ice cream or yoghurt with fruit or other flavours

## Other calcium rich meal and snack ideas

- Include canned salmon or sardines (don't forget the bones) on sandwiches, in salads or in pasta and rice dishes
- Use tofu in stir fries, pastas and rice or try it fried or scrambled as a breakfast meal
- Toss green Chinese vegetables into salads, stir fries, rice and pasta dishes and include on sandwiches

## 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. Try to be accurate in estimating amounts – measure how much soy drink is added to cereal and don't include any left in the bowl; measure how much a favourite mug or glass holds; check nutrition panels for calcium content of foods.

### 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 eg bok choy, Chinese flowering cabbage, spinach	70mg / 100g raw		
Calcium supplement	Check label		
<b>Other</b>			
		<b>TOTAL</b>	

### 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 <small>(NHMRC 2006)</small>
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

## 11. GALACTOSAEMIA AND OVARIAN FUNCTION

Galactosaemia is often associated with failure of the ovaries to develop or function properly, which can cause problems for girls during puberty or later. Most girls with galactosaemia will eventually have ovarian problems. This topic should be discussed in detail with your medical specialist.

### WHY IS OVARIAN FAILURE A PROBLEM?

The ovaries produce hormones that are associated with growth and development during puberty, and later on with fertility. The usual age at which puberty is expected to start varies between around 10-13 years for girls. This is associated with increasing growth rate and the physical changes of breast and pubic hair development. Periods usually start between 11 and 14 years of age.

If the ovaries are severely affected, the growth rate of some girls with galactosaemia may slow down at about 9 years of age and they fail to go through the normal physical development associated with puberty. Other girls progress into and through puberty normally but the ovaries gradually cease to work over several years. This may mean that the physical changes of puberty stop or that if periods have commenced, these later become irregular and gradually cease.

### HOW CAN OVARIAN FAILURE BE DIAGNOSED?

A slower gain in height than expected pre-puberty may be a useful guide to future ovarian problems. A blood test to measure levels of two hormones produced by the pituitary gland - follicle stimulating hormone (FSH) and luteinizing hormone (LH) - can sometimes predict later ovarian failure. This test is ideally done between 9 and 11 years. Unusually high levels of these hormones predict ovarian failure and will alert your doctor to make sure your child is assessed by an endocrinologist so that if necessary, some hormones can be given to ensure normal growth and pubertal development.

### WHAT CAN BE DONE ABOUT OVARIAN FAILURE?

If a girl does not enter puberty, very small doses of oestrogen are given to allow a normal growth pattern and normal physical development. This allows puberty to progress over the usual 3 years. Later, the treatment is usually simplified with a combined oestrogen and progestogen medication, used to establish regular periods. Similar treatment can be provided if ovarian failure occurs later in the teenage years but the doses of medication are increased at a faster rate in the older girl.

Some women with galactosaemia are fertile and do have babies. However, severely reduced fertility is likely if the ovaries are not functioning properly. If a young woman with galactosaemia is infertile, pregnancy can be started by obtaining an egg from another person (donor ovum), using in vitro fertilisation (IVF) to fertilise the egg with the partner's sperm and then implanting the fertilised egg into the woman's womb. A normal pregnancy can then progress although some hormone therapy will be necessary in the early stages.

In galactosaemia the scarring process in the ovaries is slow and some ova (eggs) remain present in the ovaries in childhood and adolescence. It is possible to harvest or take these eggs with a small surgical procedure and to freeze them for future use. At this time, the freezing process is satisfactory, but the thawing process is less successful. Successful use of unfertilized ova to achieve a pregnancy is extremely rare. However techniques are likely to improve and this may be a reasonable possibility for girls with galactosaemia in the future.

Contraception is a difficult issue for girls with ovarian failure. We can never be entirely sure that an ovary that is working poorly cannot produce an egg. Therefore, although girls with any form of progressive ovarian failure are unlikely to become pregnant, it is not impossible, and contraception may be required. This needs to be discussed with your doctor.

## WHEN SHOULD OVARIAN FAILURE BE DISCUSSED WITH YOUR DAUGHTER?

People's opinions vary, but it is unwise to leave any discussion about fertility problems until after puberty. A younger girl should be given information at an appropriate level that she can understand, with time to understand and accept it. Discuss the best way to handle this with your clinic Doctor.

## HOW IS HORMONE THERAPY GIVEN AND ARE THERE OTHER BENEFITS AND RISKS?

Induction of puberty usually requires daily oestrogen tablets or plastic patches that are applied to the skin once or twice every week. Initial doses of oestrogen in a small tablet include about 15 mg of lactose per day, which is considered to be insignificant. After puberty is complete, skin patches containing oestrogen can be used but many teenagers dislike them. As normal hormone levels are very important to maintain bone quality, skin, muscle and general mood and self esteem, it may be better to choose a tablet that will be taken rather than a patch that won't be used.

Girls normally accumulate about 50% of their total bone mass during puberty. This cannot be accumulated at a later time of life. If calcium intake is inadequate and proper hormone replacement treatment is not used to take a girl through puberty, she will be at serious risk of future osteoporosis (brittle bones).

Concerns have been raised in the community about hormone replacement treatment and an increased risk of breast cancer. This risk applies to a small extent to post-menopausal women and depends on the length of time the woman is exposed to oestrogen. It is not relevant to young girls with hormone deficiency. It must be remembered that the oestrogen given to girls with galactosaemia is given to make up for the oestrogen that would normally be present if the ovaries were working properly and is simply providing the missing hormone, not giving extra. The benefits of oestrogen replacement in galactosaemia far outweigh any possible minimal risks associated with this medication.

## 12. GALACTOSAEMIA INFORMATION SHEET - FOR CAREGIVERS AND SCHOOLS

Galactosaemia is an inherited condition in which the body is unable to metabolise a sugar called galactose.

Galactosaemia is diagnosed and treated in infancy. 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 needing a diet, children with galactosaemia are like any other children. 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

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

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?

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

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:

## 13. 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 heredity, 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 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, 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 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.