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GALACTOSAEMIA: SUITABLE CHEESE FOR PEOPLE ON A LOW GALACTOSE DIET



Galactosaemia is an inherited metabolic disorder of carbohydrate metabolism

which affects one in 44,000 people in the UK population, although it is more common in the Irish and Irish traveller populations. It is one of the more common metabolic disorders managed by dietitians.

Ideally, all patients should be under a specialist inherited metabolic care team, but some patients are treated in both Paediatric and Adult general district hospitals.

Classical galactosaemia is due to the deficiency of galactose-1-phosphate uridylyl transferase (GALT). Early diagnosis and treatment is vital in order to prevent liver disease and sepsis, although long-term complications of the disease do occur. In the UK, there is no screening for galactosaemia so infants are diagnosed clinically and this usually happens in the first few days of life following the development of symptoms.

Much is still unknown about this condition and its long-term outcomes. Some early treated patients develop learning difficulties. These are often mild, but can occasionally be more serious with other neurological impairments. Reduced bone density and ovarian dysfunction in females are commonly observed in older patients. Cataracts occur in untreated patients or patient non adherent to diet therapy.

MANAGEMENT

A galactose restricted diet is the only treatment for galactosaemia. The main source of galactose is from the lactose in milk, so the diet is primarily a lactose-free, galactose restricted diet. Fruit and vegetables provide a very small

amount of galactose and contribute very little to overall galactose intake, so it is not recommended that they are restricted in a diet for galactosaemia. New international guidelines on all aspects of management should be available in 2016.

Although milk-containing products are not allowed in galactosaemia, recent research has shown that certain types of mature hard cheese are low in galactose and lactose and so are suitable in this diet.

WHY SHOULD CHEESE BE ALLOWED IN GALACTOSAEMIA?

It is important that people with galactosaemia include permitted cheeses in their diet, not only for dietary freedom and enjoyment of the product, but as a source of calcium. Calcium intakes of people with galactosaemia are commonly sub-optimal despite the use of fortified products, and low intake may contribute to poor bone density. Between 2000 and 2015, the Galactosaemia Support Group has sponsored the analysis of 181 samples of cheese for lactose or for lactose and galactose, using 12 different analyses, and now has considerable knowledge about which cheese can be allowed.

Many countries now allow specific mature cheese, although these may vary from country to country.

HOW IS CHEESE MADE?

Pasteurised milk has specific starter culture bacteria added to it, so fermentation starts; then rennet is added to the ferment which causes a separation of curds from whey. The whey is a liquid which drains off and this component of the fermented milk is the part highest in lactose. Consequently, when the whey is removed, much lactose is removed with it.

When Cheddar cheese is made, further manipulation of the curds is done by salting and then ‘cheddaring’ the curds (this means cutting it into cubes and turning it constantly to dry out the cheese and encourage further whey to drain off). Traditionally, Cheddar cheese is formed into truckles (cylinders) for maturing (preferably in a cave near the Cheddar gorge).

Other cheeses such as Emmental, Gruyere and Parmesan are washed in a salt bath and this may encourage further drying out and loss of lactose.

HOW IS LACTOSE (AND THEREFORE GALACTOSE) LOST IN CHEESE MAKING?

Lactose is lost in two ways:

- 1) Lactose is metabolised by the bacteria. Some cheese have very specific bacteria added, such as propionibacter shermani in emmental cheese, which along with other strains of bacteria, make the holes. Emmental cheese is particularly

suitable in galactosaemia as the lactose is thought to be completely used by the bacteria.

- 2) Lactose is leached away from the cheese as it dries and matures, as the whey component runs off. Generally an older, more mature cheese will have less lactose as it has dried out. A soft cheese will have more moisture in it, and so more whey, and therefore will not be suitable.

WHAT ARE THE PERMITTED LACTOSE AND GALACTOSE LEVELS?

Based on many years’ experience of cheese analysis in the UK, the Galactosaemia Support Group MAP allows the use of specific cheese in galactosaemia that consistently show levels of <10mg/100g of galactose and lactose. Cheese should be analysed five times or more to allow for individual variation.

WHAT CHEESES HAVE BEEN ANALYSED?

Many different cheese have been analysed and Table 1 lists suitable and unsuitable cheese. Various milk-free cheese substitutes made predominantly for the vegan/allergy market can also be used and are free of galactose. The eating, cooking and nutritional qualities of a cheese substitute is different to a standard cheese, but they can add extra variety in the diet.

Table 1: Cheese allowed in a low galactose diet

CHEESE	DESCRIPTION	PRODUCT
Emmental cheese Grated, sliced, block	Hard Swiss cheese with holes	
Gruyere cheese Block	Hard Swiss cheese	
Comte Block	Hard French mountain cheese	

Table 1: Cheese allowed in a low galactose diet (continued)

<p>Jarlsberg Cheese Block</p>	<p>Hard Norwegian cheese with holes</p>	
<p>Italian Parmesan cheese Grana Padano, block or grated</p>	<p>Hard Italian cheese</p>	
<p>Emmi Fondue Mix</p>	<p>Swiss cheese fondue mix</p>	
<p>Specific Cheddar Cheese(UK) <i>Lye Cross extra mature and mature</i> <i>Valley Spire Parkham</i> <i>Tesco Country Farmhouse extra mature</i> <i>Sainsbury`s TTD West country extra mature</i> <i>Dromona vintage cheddar (N Ireland)</i> <i>Rathdaragh vintage cheddar (N Ireland)</i></p>	<p>Mainly extra mature or vintage cheddar block, traditionally made and packed</p>	
<p>Vegetarian/vegan cheese alternatives</p>		
<p>Bute Island</p>	<p>Sheese - Various soy cheese-hard, cream, slices</p>	
<p>Cheezy</p>	<p>Various soy cheese - hard, cream, slices</p>	
<p>Free and Easy</p>	<p>Cheese flavour sauce mix</p>	
<p>Mozzarisella</p>	<p>Creamy Risella.- Mozzarella rice based cream</p>	

Table 1: Cheese allowed in a low galactose diet (continued)

CHEESE	DESCRIPTION	PRODUCT
Vegetarian/vegan cheese alternatives (continued)		
No Muh	Cheese squares, melty cheese, herb cheese and cheese with walnuts.	
Parmazano	Grated replacement Parmesan.	
Pure	Thick Cheese Slices Pure Soft and Creamy Spread	
Tofutti	Various soy cheese - hard, cream, slices	
Vegourmet	Montanaro smoked vegan cheese slices Jeezini Celtic Cheddar style vegan cheese block Jeezy natural vegan cream cheese Bianco vegan cheese - similar to mozzarella	
Violife	Creamy Original - cream cheese type Cheddar cheese type block Parmesan	
Follow your Heart	Various cheese slices- mozzarella, herb, American cream cheese	

WHY IS SOME CHEDDAR ALLOWED BUT NOT OTHERS?

Suitable Cheddar is made using traditional methods, the right bacteria and has been matured for a long period. Traditional manufacture allows the cheese to dry naturally. Large scale factory processes pack the cheese in plastic at an earlier stage and then lactose cannot be lost.

Suitable cheese will often be labelled as extra mature or vintage Cheddar cheese and may be 12 months or 15 months old or more. Seven Cheddars are currently allowed. Lye Cross Farm cheese is still suitable, but is no longer available at Aldi.

WHY THE PDO SEAL IS NO LONGER A GUIDE TO SUITABILITY

The seal is a geographical seal, which means that the product is made in a certain area of the country. In the past, this seal was used by the GSG society to help identify suitable low lactose Cheddar cheese made in one area in Britain. However, this has caused confusion as this seal is also used for other cheese that contains lactose, e.g. Stilton has the seal as it is made near Stilton in Leicestershire, but it contains lactose. We no longer recommend using this seal to identify if a product is low in lactose and galactose.



Table 2: What is not allowed? All other cheese and cheese in manufactured foods

BabyBel - Original and Emmental
Cheddar cheeses - all except for the seven listed as permitted
Cheshire
Cottage Cheese
Dairylea Cheese Spread
Edam
Gouda
Lacto-free cheese
Processed cheese
Soft cheese
Stilton,
Wensleydale
American Parmesan
Manchego cheese - recently analysed

LACTO-FREE CHEESE - IS IT SUITABLE?

In any type of 'lacto-free' cheese, about half of the lactose is removed by filtration and the other half is enzyme treated to break down the lactose into galactose and glucose. These products contain galactose and are unsuitable for galactosaemia.



For further patient information please visit the Galactosaemia Support Group (GSG) Website: www.galactosaemia.org/.

The work on cheese would not have been possible without the support of the GSG and we are grateful for all their support.

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