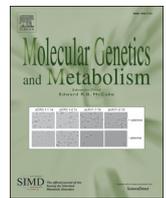




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How strict is galactose restriction in adults with galactosaemia? International practice

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ABSTRACT

Dietary management of 418 adult patients with galactosaemia (from 39 centres/12 countries) was compared. All centres advised lactose restriction, 6 restricted galactose from galactosides \pm fruits and vegetables and 12 offal. 38% (n = 15) relaxed diet by: 1) allowing traces of lactose in manufactured foods (n = 13) or 2) giving fruits, vegetables and galactosides (n = 2). Only 15% (n = 6) calculated dietary galactose. 32% of patients were lost to dietetic follow-up. In adult galactosaemia, there is limited diet relaxation.

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1. Introduction

There has been considerable debate regarding the necessity for severe galactose restriction in older patients with galactosaemia. Patient support groups are posing questions about the essentiality of life-long strict diet and some adults with galactosaemia may already follow a self-relaxed diet. Although poor dietary control beyond infancy has been associated with cataract formation [1,2] and increases in biochemical markers [3], overall life-long dietary restriction does not appear to play a significant role in determining the severity of cognitive outcomes [4–6]. In practice, there is little data about the dietary management of adult patients and many are lost to dietetic follow-up [7]. There are only a few adult case reports of patients homozygous for the severe mutation Q188R who have self-liberated galactose-intake without obvious adverse effects [8–10]. In order to assess the extent of diet relaxation in adult clinical practice, this international survey was conducted to examine dietary advice given to adults with galactosaemia.

2. Materials and methods

2.1. Study design

A questionnaire (17 multiple choice and short answer questions) relating to dietary advice to adult patients with galactosaemia, was sent to dietetic members of the Society for the Study of Inborn Errors of Metabolism (SSIEM) and Australasian Society for Inborn Errors of Metabolism (ASIEM). Dietitians were requested to cascade this questionnaire to other dietetic colleagues within their country. In this cross-sectional audit, data from each clinic was collected about the severity of dietary restriction (quantity of galactose allowed; restriction of galactosides, galactose in fruit and vegetables, offal; and cheese permitted), any dietary changes by patient or dietitian specifically in adulthood, circumstances when dietary treatment might be relaxed, regularity of follow-up; and frequency of galactose-1-phosphate (Gal-1-P) monitoring. Data on patient genotype, long term patient outcome and biochemical data was not included in this audit. Ethical approval was not required as no specific identifiable patient data was obtained.

3. Results

Questionnaires were returned from 39 European and Australasian international IMD centres providing data on 418 adult patients with galactosaemia from 12 countries (Table 1). Each individual centre regularly treated between 1 and 24 adult patients (median: 8 patients) with galactosaemia.

3.1. Overall dietary restrictions

All centres recommended lactose restriction in adult patients, but there was some galactose restriction from fruits and vegetables (4/39)

and galactosides (6/39) and 12/39 centres limited offal intake (Table 1). Only one centre in Spain did not permit any low lactose cheese.

3.2. Relaxed diets

In adults, diets were relaxed in 15 centres (38%) from 6 countries (Table 1), but mainly self-directed by patients than dietitians. Centres advising a lactose-free diet only, relaxed diets with trace amounts of lactose in manufactured foods such as breads, biscuits, and cakes (e.g., lactose in sodium and calcium caseinates, animal fats, flavourings) (estimated galactose: <20 mg/portion) or margarine and butter on sandwiches (estimated galactose: up to 200 mg/sandwich) purchased in retail outlets. Centres in France permitted all fruits, vegetables and galactosides and so advised a lactose-free diet only. Although information was not collected about metabolic control, no dietitians reported development of symptoms as a consequence of these small changes.

3.3. Calculation of galactose intake

Most centres (85%, n = 33) did not formally calculate the amount of galactose in the diet. One centre (Belgium) allowed only 50 mg/day, 2 centres (Italy, Netherlands) 100 mg/day, 1 centre (Belgium) 300 mg/day, 1 centre (Germany) 300–500 mg/day, and 1 centre (Germany) 500 mg/day.

3.5. Follow-up

Patients were monitored by dietitians annually by the majority of centres (67%, n = 26). Thirty-two percent (n = 132) of patients were lost to dietetic follow-up (definition: no contact for at least 2 years). Reasons for lack of follow-up included: patient relocation, regular follow-up by physicians but not dietitians, and patient choice only to attend clinics according to need e.g. pregnancy and genetic counselling.

Gal-1-P measurements were monitored by half of the centres (n = 20) annually and 14 (36%) rarely or never monitored this biochemical marker.

4. Discussion

This is the first international multi-centre study to describe dietetic management practices in adult patients with galactosaemia. It is clear that there is uncertainty regarding the requirement for strict diet in adulthood. Although over 75% of centres considered that a strict diet was necessary, lower patient dietary adherence associated with less health professional conviction for the need for strict diet led to a very cautious approach in moderating dietary restriction. Advice in adulthood was also influenced by severity of childhood restriction (lactose free only vs low galactose). To our knowledge no adult suffered from symptoms as a consequence of small dietary changes but there was

Table 1
Dietary treatment policy and diet relaxation practices for adults with galactosaemia reported by centres.

Country represented by participating centres	No. of centres from each country	Total number of patients	Dietary treatment policy – restricted foods			Allow relaxed diet	Reasons for diet relaxation			Types of foods permitted in relaxed diet	
			Lactose	Galactose from fruit and vegetables	Galactose from galactosides		Galactose from galactosides	Galactose restricted from offal	Patient request		Dietitian does not consider strict diet necessary
UK	11	109	11/11	0/11	0/11	0/11	4/11	1/1	4/11	0	Lactose containing ingredients in manufactured foods
Germany	6	65	6/6	0/6	4/6	2/6	1/6	1/6	1/6	1/6 centres in patients >14 years of age	Lactose containing ingredients in manufactured foods
Netherlands	5	51	5/5	0/5	0/5	3/5	2/5	3/5	1/5	0	Lactose containing ingredients in manufactured foods
Belgium	4	21	4/4	1/4	1/4	3/4	0	2/4	2/4	0	3/4 centres lactose containing ingredients in manufactured foods
Australia	2	54	2/2	0/2	1/2	0/2	1/2	0	0	0	None
France	2	21	2/2	0/2	0/2	2/2	0	1/2	2/2	2/2 in centres in patients >10 years of age	Diet relaxed to a lactose-free diet only
Italy	2	19	2/2	2/2	2/2	0/2	0	0	0	0	None
Spain	2	16	2/2	1/2	2/2	0/2	0	0	0	0	None
Sweden	2	11	2/2	0/2	1/2	0/2	0	0	0	0	None
New Zealand	1	44	1/1	0/1	0/1	1/1	1/1	1/1	1/1	0	Lactose containing ingredients in manufactured foods
Norway	1	4	1/1	0/1	0/1	0/1	0	0	0	0	None
Switzerland	1	3	1/1	0/1	1/1	0/1	0	0	0	0	None
TOTAL	39	418	39/39 (100%)	4/39 (10%)	6/39 (15%)	15/39 (38%)	9/39 (23%)	9/39 (23%)	11/39 (28%)	3/39 (8%)	Lactose containing ingredients in manufactured foods

Definitions: lactose restriction: avoidances of animal milk and products contains animal milk e.g. yoghurt, ice cream, lactose in manufactured foods; galactose-restriction from fruit and vegetables: avoidance of high galactose containing fruit/vegetables e.g. tomatoes, kiwi fruits, banana; avoidance of galactosides e.g. peas, beans, lentils, cocoa; restriction of galactose storage organs e.g. offal: liver, kidney, and eggs.

limited monitoring of biochemical control. It was disturbing that almost a third of patients were lost to dietetic follow-up, and some were reviewed by a medical physician without dietetic follow-up. Treatment centres should strive to maintain long-term dietetic follow-up of all patients.

Disappointingly, there was still rigorous restriction of galactose extending beyond lactose restriction (elimination of galactose from some fruit and vegetables, galactosides and galactose storage organs) by a small number of centres. There is little evidence to support this practice, and a recent USA review recommended that fruits, vegetables, legumes and non-fermented soya products are given without restriction in a low galactose diet [11] to all age groups.

In conclusion, almost 40% of IMD centres relax diet in classical galactosaemia, but any change is commonly patient led and small (<200 mg/day galactose). Additional galactose intake was not usually quantified, generally hampered by a lack of reliable data on the galactose content of foods. Although it is essential that further prospective multi-centre research examining individual galactose tolerance is conducted before recommendation on diet relaxation is given, health professionals should report the outcome of individual adult patients on less restrictive diets, and any galactose intake should be systematically estimated at clinic review.

Conflicts of interest

Sarah Adam – funding from SHS and Vitaflo to attend study days and conferences.

Rhonda Akroyd – funding from SHS and Vitaflo to attend study days and conferences.

Alberto Burlina – honoraria from Nutricia, and Merck Serono, Member of Sapropterin Advisory Board (Merck Serono international), Member of the Advisory Board Element (Danone-Nutricia).

Karen Corthouts – support from Nutricia, Vitaflo, Merck Serono to attend independent or industry-organised trainings and conferences.

Marianne Diels – support from Nutricia, Vitaflo, Merck Serono to attend independent or industry-organised trainings and conferences.

Katharina Dokoupil – member of the European Nutrition Expert Panel (Merck Serono international).

Sharon Evans – a research dietitian funded by Nutricia; financial support from Nutricia and Vitaflo to attend study days and conferences.

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Pat Portnoi – sponsorship from Nutricia and Vitaflo to attend conferences and study days.

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Marine Robert – member of the European Nutrition Expert Panel (Merck Serono International).

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Linn Helene Stolen – travel/accommodation/meeting expenses from Nutricia and Vitaflo.

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Authors' contributions

All authors were involved in data collection, interpretation of data, critical revision of the paper for important intellectual content and final approval of the version to be published. Anita MacDonald and Pat Portnoi were additionally involved in the initial conception and design and Anita MacDonald and Sharon Evans in the collation of data and drafting of the initial article.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <http://dx.doi.org/10.1016/j.ymgme.2015.03.008>.

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