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Dietary information for the management of Adult Refsum disease

What is Adult Refsum disease?

Adult Refsum disease is a rare condition affecting approximately one in a million people. People are born with this condition because they inherited a faulty gene from both of their parents. The gene affects the body's ability to process a type of fat found in the diet called phytanic acid. Phytanic acid builds up in the body from eating foods high in phytanic acid, or from situations that cause the body to break down fat stores, releasing phytanic acid, including fasting, illness, low carbohydrate diets, exercise without appropriate fuel, weight loss and high caffeine intake.

The levels of phytanic acid build up in the body over time and eventually cause symptoms of worsening vision, hearing and sense of smell. There can be a loss of sensation in the feet and legs. There may be difficulty in coordinating walking and balance. The skin can become itchy and scaly as well as other symptoms.

What is the treatment for Adult Refsum disease?

Treatment aims to reduce the phytanic acid content of the blood through diet and, if necessary, by a treatment called plasmapheresis. When the phytanic acid content in the blood falls, there can be a return of sensation to the feet and legs, improvements in walking and balance and the skin can return to a healthy state. Damage to the eyes, hearing and sense of smell cannot currently be reversed, but further loss is generally slow to occur.

How does the dietary management of Adult Refsum Disease work?

The Refsum's Diet has been shown to be effective at lowering the amount of phytanic acid in the blood. The diet works by:

1. Restricting foods high in phytanic acid.
2. Reducing the release of phytanic acid from body stores with having regular meals containing carbohydrates, getting enough energy (calories), fuelling appropriate around exercise, managing illness with an emergency regimen, preventing unplanned weight loss and limiting caffeine.

How quickly will my phytanic acid fall?

The phytanic acid content of the blood will gradually decrease over time. The decrease is not normally a straight line and the rate of decline varies from person to person.

Where does phytanic acid come from?

Phytanic acid is present in larger amounts when phytol, which is part of the chlorophyll molecule, is fermented by bacteria. This occurs in the stomach of ruminant animals (cows, sheep).

There are also two substances that can be converted to phytanic acid by the body: phytol fatty acid esters and free phytol. Both of these substances come from plant-based foods such as vegetables, fruit, nuts, seeds and legumes.

How is this diet guidance different from the 2024 version?

Global DARE Foundation funds the analysis of foods which informs the updates to the guidelines. This 2026 guidance contains a number of foods, which have recently been analysed. The structure of the diet guide, using the traffic light system to categorise foods, remains the same. The

diet guide continues to include Level 1 and Level 2 advice. We know that phytanic acid in foods directly affects phytanic acid blood levels. However, further research is still needed to understand how phytyl fatty acid esters and free phytol are absorbed in the gut and how they affect phytanic acid levels in the body. Until we have this research, it has been decided that the Level 1 advice is based only on phytanic acid content of foods. In addition, Level 1 advice includes recommendations for regular meals, carbohydrates, exercise and illness management.

Level 2 advice considers foods high in phytyl fatty acid esters that could potentially influence phytanic acid blood levels. Current food analysis does not provide conclusive free phytol content, and therefore, it has not been considered in Level 2 diet guide until we have further research.

What level of dietary advice should I be following?

We would recommend following all the Level 1 advice as a first line in managing your phytanic acid levels. If, after following all the Level 1 advice, you continue to have higher phytanic acid levels, we would recommend also considering the Level 2 guidance (in addition to the Level 1 advice).

Important information: Level 1 and Level 2 assess different substances in food. A food may be low in phytanic acid and therefore suitable under Level 1, but contain higher amounts of phytyl fatty acid esters and therefore be restricted under Level 2. If you have been advised to follow Level 2, and there is a food that is listed as being able to eat under Level 1 guidance but is now restricted under Level 2, the Level 2 guidance takes priority for that food item.

Level 1 guidance

It is recommended that all people with Adult Refsum disease follow the Level 1 dietary recommendations initially.

Please note, the guidance below provides general dietary recommendations that can be used by all individuals with Adult Refsum disease. However, everyone's food choices, portion sizes and phytanic acid levels are different, so it is recommended to speak with your dietitian to get individualised advice.

What foods can I safely eat?

These foods contain only small amounts of phytanic acid and can be eaten ***freely***.

Meats and eggs

Chicken
Chicken liver
Duck breast
Eggs
Turkey
Lean pork (cuts such as pork tenderloin and loin)
Low fat ham
Bacon (back cut or loin bacon only with fat removed)

New additions:

Dried pork (prosciutto)
Salami (pork only)
Chorizo (pork only)
Wild boar (wild pig)

Seafood

Clams
Coley
Lobster (Maine)
Perch (fresh water)
Prawns / shrimp
Scallops (bay and deep sea)

New additions:

Sole (dover and lemon)

Meat alternatives

Quorn

Tofu

New additions:

Beyond burger – *(note this has been analysed on a US specific product. If consumed outside of the US, please check the ingredients list does not contain any products that are listed as needing to be avoided in this diet guide).*

Legumes

Black beans

Butter beans/ lima beans

Chickpeas

Edamame beans

Green lentils

Red kidney beans

New additions:

Cannellini Beans (white kidney beans)

Soya beans

Black eyed beans/ peas

Gigantes (Greek white beans)

Borlotti/ cranberry beans

Pinto beans

Nuts and seeds

Almonds

Black chia seeds

Black sesame seeds

Brazil nuts

Cashew nuts

Coconut

Golden flaxseed

Hazelnuts

Macadamia nuts

Peanuts

Pecans

Pine nuts

Pistachios

Pumpkin seeds

Sunflower seeds

Tahini (white sesame seed paste)

Walnuts
White sesame seeds
Nut butters made from any of the above nuts/seeds

Milk and milk alternatives

Coconut, oat, and soya yoghurt
Fat-free cow's yoghurt
Fat free or 1-2% fat cow's milk (with no DHA)
Oat, coconut, rice and soya milk
Very-low fat fromage frais
Whey powder
Dairy free whipping cream

Cheese and cheese alternatives

Vegan and dairy-free cheese
Vegan cheese spread
Fat-free mozzarella

Cereal and cereal products

Bread (white, rye, wholemeal, wholegrain)
Breakfast cereals (with no added milk/dairy)
Crumpets
Flour (white, wholemeal)
Pasta / noodles
Pearl barley
Porridge oats
Rice
Sago
Tapioca

New additions:

Quinoa (white)

Fruits

All fresh and dried fruit

Vegetables

All fresh and frozen vegetables

Fats and oils

Vegetable oils

Nut and seed oils made from any of the nuts and seeds listed above
(except flaxseed / linseed oil)

Vegetable suet

Vegan and vegetable oil spreads

Miscellaneous

Nori

Wasabi

Fish sauce

Herbs

Spices

Ketchup / tomato sauce

Bisto gravy powders

Stock - chicken or vegetable

Honey

Jam

Marmite

Cocoa powder

Crisps / potato chips

Ready-made sauces containing low phytanic acid ingredients (including whey protein, milk protein or skimmed milk)

Homemade puddings with only low phytanic acid ingredients

New additions:

Gelatine (fish and bovine; Note: vegetable gelatine alternatives have not been tested)

Cacao butter

Biscuits and confectionery

Boiled sweets or sugar candy

Dairy free baking (not containing any dairy milk, butter and cream)

Dark chocolate (e.g. 70 or 90%)

Liquorice

Vegan / dairy-free chocolates

Vegan baking

Vegan ice cream

Beverages

Tea

Fruit juice

Coffee (see information on caffeine recommendations)

CAUTION

These foods need to be **limited** as they contain moderate amounts of phytanic acid.

A total of a **100g portion** of one or any combination of the below foods can be taken **once a day**.

If your phytanic acid levels are not responding to these restrictions (i.e., remain high), consult your dietitian for specific guidance.

Meats

Frankfurter sausage

Rabbit

Veal

Fatty bacon

Pork belly, pork lard, pork sausages, and pork shoulder (higher fat cuts)

Mortadella

Seafood

Calamari (not coated/crumbed)

Halibut

Herring

Tinned tuna (in water / brine)

New additions:

Monkfish

Legumes

Baked beans

Dairy

Fat-free and low-fat cottage cheese

Cereals

Rice crispies

STOP

These foods need to be ***avoided*** as they are high in phytanic acid.

Meats

Beef
Goat
Lamb
Mutton
Sheep
Venison
Pork offal (tongue, liver, kidney)

Seafood

Cod
Crab
Flounder
Fresh tuna
Haddock
Mackerel
Oysters
Plaice
Salmon
Sardines
Smoked salmon
Trout

New additions:

Pollock
Sea bass
Whiting
Pike

Milk and milk products

All dairy creams (e.g. double cream, sour cream, single cream, whipping cream)
Buttermilk
Fat-free milk with added DHA (Omega 3)
Full-fat, whole or milk with more than 2g fat per 100ml
Evaporated milk
Full-fat yoghurt
Ice cream

Cheese and cheese products

Cheese spread

Full fat cottage cheese

Cream cheese

Hard and soft cheeses (cow, sheep, goat, including lite cheese)

Processed cheese

Halloumi

Cereal and cereal products

Breads with dairy (e.g. brioche, croissants)

Cereals with dairy

Vegetables

Potato flakes, dried potato or packet mash potato (if dairy / milk present in ingredients list)

Fats and oils

Algae oil

Beef suet

Butter and half-fat butter

Flaxseed / linseed oil

Fish oil

Ghee

Margarine (if it contains milk and milk products)

New additions:

Krill oil

Miscellaneous

Stock – beef or lamb

Ready-made sauces containing dairy such as cream or milk

Biscuits and confectionery

All baking that includes butter, milk, or other dairy

Chocolate (white and milk)

Carbohydrates are important: These should be included at each meal

The brain needs a regular supply of glucose to function. Without a regular supply of glucose, your body will break down its carbohydrate stores and once it has used those, it will break down your fat stores and release phytanic acid from your body fat. It is very important **not** to follow a low carbohydrate diet. Carbohydrates can come from starchy foods (bread, potatoes, rice, pasta, breakfast cereal) and sugary foods (fruit, sweets, fruit juice). Carbohydrates should be distributed throughout the day (included at each meal and in snacks, including a bedtime snack), and should not be less than 200g of carbohydrates in total per day. Carbohydrate free meals should be avoided.

If you have other health conditions such as diabetes, it is important to talk with a dietitian for specific advice.

Ensuring adequate carbohydrates when exercising

It is important to fuel your body with carbohydrates prior to exercise. This could include eating a carbohydrate containing meal or snack before exercising. If exercise lasts longer than 45 minutes, you should take a carbohydrate snack during exercise. For example, if you go on a 90-minute walk you should bring a small snack to eat at 45 minutes (e.g. banana, a couple of dates). If you exercise without adequate carbohydrates, your body will start to break down fat and release phytanic acid. If you are starting a new exercise plan or wanting to increase the amount of exercise you do, it is important to talk with your dietitian.

Protein sources for Adult Refsum disease

Protein is an important nutrient for growth, development and maintenance of muscle. Protein foods should make up one-third of your plate at meals. Some protein foods are very high in phytanic acid (e.g. beef, lamb, cheese, full-fat milk) and need to be avoided. Below is a list of protein foods that can be eaten daily. You will also get some protein from vegetables, cereals and grains. You can use the traffic light lists above for

further ideas of protein foods (including those that can be eaten in moderation on the 'caution' list).

- Chicken, turkey, duck breast, lean pork eg pork loin, ham without fat, back bacon without fat
- Eggs
- Prawns/shrimp, scallops, lobster, clams
- Perch (fresh water), coley, sole
- Quorn
- Tofu
- Kidney beans, black beans, green lentils, butter beans, chickpeas
- Whey powder
- Fat-free cow's yoghurt
- Soya yoghurt
- Fat-free milk
- Almonds, cashew nuts, walnuts, pistachios, hazelnuts, macadamia nuts
- Pumpkin and sunflower seeds

Other Level 1 dietary recommendations:

Eat regular meals and snacks

This means having breakfast, lunch, evening meal and a bedtime snack. If you have a long break between meals (4 hours or more), have a snack. It is particularly important to eat breakfast, so that you break your overnight fast. It is also important that you do not exercise in the morning unless you have eaten breakfast.

New information: If your appetite is low, it is important to raise this with your Refsums team so they can support you to get enough calories through food and/or oral nutritional supplements. If oral nutritional supplements are required, it is important to check it does not contain any ingredients that need to be avoided such as milk, fish oils or peanut oil.

Whey protein or milk protein are commonly used in these products, but they only contain the protein portion of milk and are therefore safe to consume.

Dieting to lose weight is **not** recommended unless under close supervision of your medical and dietitian team. If weight loss is desired, it is important that you work closely with your medical team and dietitian to develop a weight loss plan and regularly monitor your phytanic acid levels. Any time you lose weight phytanic acid is released from your fat stores; therefore, very slow weight loss would be recommended.

Avoid high intakes of caffeine

Consuming large amounts of caffeine from energy drinks, sports drinks, or high intake of coffee is not recommended. If consuming products with caffeine, a moderate caffeine intake is advised. For an adult woman (not intending to become pregnant) this would be 200-350mg a day and for an adult man, 300-450mg a day. Examples of caffeine content in various products are listed below:

Product	Serving Size	Caffeine Content
Sports or energy drinks	Per can or bottle	80-300mg
Mug of tea	250mls	40-70mg
Mug of instant coffee	250mls	52-85mg
Cola	330ml can	11-70mg
Pharmaceutical products	Per tablet	25-65mg
Energy supplements	Per supplement	35-50mg

Getting enough glucose when unwell

If you are feeling unwell and unable to manage all your usual meals, you will need to consume glucose at regular intervals in order to stop the breakdown of your fat stores and release of phytanic acid into your body. This is called an emergency regime, and an example of one used at Guy's

and St Thomas' Hospital, London, is included (see Emergency Regimen). This regimen is specific for those who do not have diabetes. If you have diabetes, it is important to contact your medical team and dietitian for specific advice.

Medical procedures or surgery that requires fasting

Fasting for a surgical or medical procedure can result in increasing phytanic acid levels. To prevent the increase in phytanic acid levels, glucose solutions (oral and IV) will need to be provided prior, during and post procedure. If you are having a surgery or medical procedure, please contact your medical/dietetic team to make them aware.

Ensure you are getting your essential fatty acids

Omega 3 and omega 6 are essential fatty acids that the body cannot synthesise and therefore need to be consumed in the diet. Omega 3 and 6 are found in vegetable oils, and can be a source of essential fatty acids on a low phytanic acid diet. In addition, omega 6 can also be found in chicken, eggs and wholegrain breads. It is important to discuss with your dietitian about ensuring you are getting omega 3 and 6 from your diet.

Ensure adequate vitamins and minerals

With all specialist diets it is important to regularly monitor nutritional bloods (for example, vitamin B12, folate, ferritin, vitamin D). These should be monitored annually and vitamin / mineral replacement provided as needed.

Level 2 guidance

If phytanic acid levels continue to be high despite following **all** Level 1 advice, we would recommend reviewing your diet to see if any of the below foods feature. These foods contain phytyl fatty acid esters, which could be contributing to your phytanic acid levels in your blood. If you are consuming any of these foods regularly, it is recommended to speak with your dietitian for specific guidance.

CAUTION

These foods contain **moderate amounts** of phytyl fatty acid esters.

A total of a **100g portion** of one or any combination of the below foods can be taken **once a day**.

Tahini (white sesame seed paste)

White sesame seeds

Pumpkin seeds

Sesame seed oil

Olive oil

Soya oil

Peanut oil

Peanut butter

Peanuts

Pecans

Broccoli

Rocket / arugula

Red and yellow bell peppers

Green olives

Edamame beans

Chickpeas

Dairy free / vegan butters, cheese, cream cheese, cheese spread or whipping creams **containing any of the above oils**

STOP

These foods are **high** in phytol fatty acid esters.

Flaxseed / linseed oil

Black sesame seeds

Sunflower seeds

Golden flaxseed

Kale

Beyond beef

Dark chocolate

Dairy free / vegan butters, cheese, cream cheese, cheese spread or whipping creams **containing flaxseed / linseed oil**

New additions:

Gigantes (Greek white beans)

Cannellini Beans (white kidney beans)

Cacao/ cocoa butter

Vegan/ dairy free chocolate containing cacao/ cocoa butter

Meal swap examples

Level 1 guidance



Croissant, butter and coffee

× High phytanic acid foods



Wholemeal bread, vegan spread, eggs, orange juice

✓ Low phytanic acid foods

Description: A breakfast with croissant, butter and coffee that would be high in phytanic acid. This can be swapped for two slices of wholemeal bread with vegan spread, eggs and glass of orange juice as a breakfast lower in phytanic acid.

Level 1 guidance



1 x bread, vegan spread and tea

× Not enough calories



2 x bread, vegan spread, soya yoghurt, banana and tea

✓ Adequate calorie

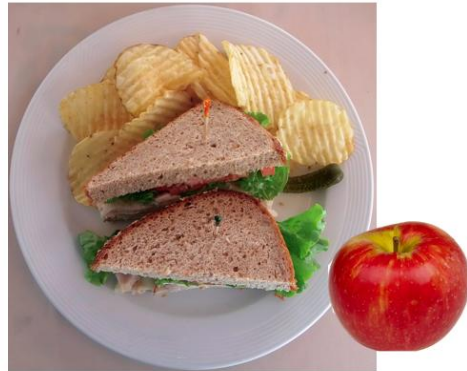
Description: A meal of one slices of bread with vegan spread and cup of tea. This meal would not contain enough calories. To ensure adequate calories the meal could be swapped for two slices of bread with vegan spread, along with soya yoghurt, a banana and tea.

Level 1 and 2 guidance



Chicken, rocket/arugula, carrots, apple

- × Low in carbohydrates
- × Moderately high in phytol esters



Chicken salad sandwich (wholemeal bread, lettuce, vegan spread, tomato), crisps and apple

- ✓ Carbohydrate portion
- ✓ Low in phytol esters

Description: A lunch of grilled chicken, rocket/arugula, carrots salad and an apple. This lunch would be too low in carbohydrates (Level 1 advice) and contain foods moderately high in phytol esters (Level 2 advice). This can be swapped for a chicken salad sandwich, crisps and an apple, which would contain a carbohydrate portion and be lower in phytol esters.

Emergency regime

Example of regimen used at Guy's and St Thomas Hospital, London

The emergency regime (ER) of glucose polymer drinks should be commenced immediately if you become unwell, e.g. nausea, vomiting, diarrhoea, high temperature, or any illness resulting in loss of appetite and inability to take your normal diet.

The ER is used to prevent the release phytanic acid from your body fat which can occur if your body is not getting adequate carbohydrates during illness. It is important to have the right amount of glucose to maintain your body metabolism, and prevent breaking down of your body's alternative source of energy during this time.

Stage 1

At the first sign of feeling unwell and loss of appetite, take 200 ml glucose polymer drink (**see recipes on the following page**). If you feel better within one to two hours, you may return to your normal diet.

Stage 2

If you continue to feel unwell, commence the full emergency regime as described below and contact the Inherited Metabolic team.

Stage 3

If you are unable to tolerate the emergency regime, if you do not improve, or if you become increasingly unwell, go to your local Emergency department and ask the hospital healthcare team to contact the Inherited Metabolic team.

The full emergency regime

- Take the ER drinks every two hours **day and night**
- If tolerated, continue all prescribed medicines

- You should not use your ER for more than 2 days without your normal diet. If your symptoms persist and you are unable to tolerate your ER, contact your GP or go to your nearest Accident and Emergency (A&E). Inform your Inherited Metabolic team.
- Continue with extra glucose polymer drinks during the day until the normal diet is resumed

Recipes for 20% emergency regimen drink

One of the following recipes will be recommended by your dietitian. These can be made up using 200ml of water. Consume all 200ml of the ER drink every 2 hours day and night.

- 40g Maxijul Super Soluble, Polycal or Vitajoule powder
- OR 1 sachet of S.O.S 20 (Vitaflo)

Alternative oral emergency drinks for adults

Ideally 200ml of the emergency drink (as per recipe above) must be taken every 2 hours when you are unwell. If you cannot take any of the ER drink or you don't have access to them quickly, then you can start with an alternative drink such as the options given below. You will need to drink more than 200ml of the alternative drinks every 2 hours to get your carbohydrate needs.

Mountain Dew Citrus Burst (13g carbohydrates per 100ml)	Take 300ml every 2 hours
Coca Cola (full sugar) (10.6g carbohydrates per 100ml)	Take 380ml every 2 hours
Pepsi (full sugar) (11g carbohydrates per 100ml)	Take 380ml every 2 hours
Fruit juice (eg apple juice) with at least 10g carbohydrates per 100ml	Take 400ml every 2 hours

Note:

- Carbohydrate content of the drinks can change. Please regularly check the carbohydrate content per 100ml to ensure it still contains the above amount of carbohydrate.
- Some of the above alternative drinks contain caffeine. Use of these for 24 hours during illness would be appropriate. When well, to consume in moderation.

Contact Details

St Thomas' Hospital Adult Inherited Metabolic Service

Consultant's Secretary:	gstt.adultmetabolic@nhs.net Monday-Friday (0900-1700) Tel: +44 (0) 20 7188 4004
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Patient Support Group

Global DARE Foundation

Global DARE Foundation's mission is to promote world-wide awareness and better quality of life for all who are diagnosed with Adult Refsum Disease. DARE stands for Defeat Adult Refsum Everywhere.

At DARE's website there are educational resources including recorded webinars on Refsum disease and this diet.

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