

Question on the updated Refsum Diet Guide (version January 2024)

Question 1	During the webinar, you suggested that if phytanic acid levels are well controlled, then we can confidently continue to follow the Level 1 guidance. Can you elaborate more on the meaning of «well controlled» levels.
Answer	<p>In general, the effectiveness of the Level 1 diet guidance can be assessed by observing the trend in phytanic acid (PA) levels over time. For instance, consider an individual diagnosed with adult Refsum disease two years ago, whose initial PA level was 1500 umol/L. If their levels have decreased to 800 umol/L, it indicates a positive response to the diet, suggesting that it is working effectively for them.</p> <p>Similarly, if another individual with adult Refsum disease has maintained stable PA levels around 200 umol/L or lower for an extended period, this also suggests a favorable response to the dietary regimen.</p> <p>Conversely, a lack of response to the diet would be indicated by a failure to observe any decrease in PA levels, or if the levels persist stubbornly high despite adhering to all recommended dietary guidelines.</p> <p>It's important to note that PA levels may not decline consistently over time. External factors such as illness, like the flu or gastroenteritis, can temporarily elevate PA levels, leading to fluctuations in readings. However, these fluctuations do not necessarily indicate diet ineffectiveness.</p> <p>While PA levels are not expected to decrease linearly, a graph plotting the readings should demonstrate either a gradual decline or stability at a low to moderate level, indicating the effectiveness of the dietary intervention over time.</p>

Question 2	How long would you wait to say that the Level 1 diet is not sufficient and that we should introduce the Level 2 guidance? At what level of plasma phytanic acid would you switch from Level 1 to Level 2?
Answer	Our recommendation is to focus on the overall trend of your phytanic acid levels. If you notice a gradual decrease over time, it suggests that your current approach is effective, and there's likely no need for

	<p>immediate changes. Introducing additional restrictions can unnecessarily complicate matters.</p> <p>However, if you reach a point where your levels plateau, for example, around 800 umol/L, and you're not seeing further improvement, it may be worth considering Level 2 guidance. It's important to understand that reducing phytanic acid levels, especially when they are initially high, can take considerable time. Be patient and monitor the trend closely.</p> <p>Discuss with your healthcare team the frequency of monitoring your phytanic acid levels. While weekly assessments may be excessive, checking every four to six months can provide valuable insights into the effectiveness of your dietary regimen. It's crucial to give the diet time to produce results and to make decisions based on long-term trends rather than short-term fluctuations.</p>
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Question 3	<p>Given many of us only get PA levels back infrequently, how do we know we need level 2 advice? If so, given the amount of time passed in getting PA levels measured, how can we attribute to particular foods? (I imagine this could only work if you eat certain foods and then can get a quick PA test to get results). For most of us it is going to be a guesstimate as to impact of particular foods.</p>
Answer	<p>It is common practice to have the phytanic acid test every six to twelve months. Our recommendation is to focus on the overall trend of your phytanic acid levels. If you notice a gradual decrease over time, it suggests that your current approach is effective, and there's likely no need for immediate changes. Introducing additional restrictions can unnecessarily complicate matters.</p> <p>However, if you reach a point where your levels plateau, for example, around 800 umol/L, and you're not seeing further improvement, it may be worth considering Level 2 guidance. It's important to understand that reducing phytanic acid levels, especially when they are initially high, can take considerable time. Be patient and monitor the trend closely.</p>

Question 4	Was free phytol previously considered in the diet guide? Which foods are no longer included in the diet guide because of that?
Answer	<p>Yes, the previous dietary guidelines treated phytyl fatty esters, free phytols, and phytanic acid on a one-to-one basis. Consequently, all the foods now categorized as Level 2 were restricted in the previous guidelines, except for a few items for which we have obtained new analytical data.</p> <p>Previously, all Level 1 and Level 2 foods were subject to limitations. For instance, chickpeas or edamame beans were included in the caution (amber) list because both phytanic acid and phytyl esters were combined, assuming they collectively influenced phytanic acid levels in the blood. Consequently, they were classified under the amber category. However, upon considering only phytanic acid and excluding phytyl esters, these foods have now been reclassified under the green category.</p> <p>Many foods have transitioned from amber to green, or from red to amber, and some have been newly categorized under Level 2 advice.</p> <p>Therefore, we recommend that if you are currently using a dietary guide from the previous version dated 2022, please replace it with the new one, as there have been significant changes in the categorization of foods.</p>

Question 5	Flaxseed/linseed oil is included in both Level 1 red and Level 2 red?
Answer	<p>Yes, and the reason for that is they are high in phytyl fatty esters.</p> <p>Since they're not high in phytanic acid, technically they don't fall under Level 1, which is solely based on phytanic acid content. However, their phytyl fatty ester content is significantly elevated. We felt that if someone incorporated these oils into their diet, it could potentially impact phytanic acid levels. Therefore, we've included them in the Level 1 advice, opting to err on the side of caution.</p>

Question 6	How much phytanic acid results in a product being categorized in the green, amber or red group?
Answer	<p>The categorising of foods is based on the assumption that an individual with Adult Refsum can still process around 10 milligrams of phytanic acid a day through omega-oxidation.</p> <ul style="list-style-type: none"> - Any food that is below 2 milligrams of phytanic acid per 100 grams a day is in the green category. - Any food that has between 2 and 5 milligrams of phytanic acid per 100 grams is in the amber category - Any food above 5 milligrams of phytanic acid per 100 grams a day is in the red category. <p>Depending on one's eating habits, if a food categorized in the green section happens to be higher in phytanic acid and is consumed in large quantities at every meal, it could accumulate. This underscores the importance of collaborating with your dietitian or healthcare team.</p> <p>Nevertheless, considering the typical dietary patterns and variety in people's diets, we believe that foods containing less than 2 milligrams per 100 grams should generally be safe to consume. However, those falling between 2 and 5 milligrams per 100 grams have been categorized with a recommended portion size of 100 grams per day to advise against consuming large amounts, as this could lead to significant accumulation over time.</p>

Question 7	Have I understood well that there are no nuts to be avoided anymore in Level 1 advice like there as there used to be? Are all nut levels known yet?
Answer	<p>Correct. Nuts that are on the list for Level 1 advice can be had freely. There are no nuts on that list that need to be cautioned or avoided. We haven't analysed all nuts yet, but we've got the list there in the Level 1 guidance and any of those that are included there, or their oils versions or their butter versions, such as peanut butter or almond butter can be consumed freely as part of level one advice.</p>

Question 8	Do you recommend people currently following the Refsum diet to reintroduce Level 2 foods?
Answer	<p>Assuming that levels are well controlled these foods could be introduced again. However, please have a discussion with your dietitian, your healthcare professional team as it also depends on how much you would have of those foods. It is important to monitor</p>

	your levels. But again, it is also really important to think about your dietary patterns and not be solely focused on foods in different categories. Ensure that your carbohydrate intake is sufficient, etc.
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Question 9	Would you switch to Level 2 guidance solely due to neurological symptoms regardless of phytanic acid levels?
Answer	<p>It shouldn't be an automatic decision to switch dietary plans because there are numerous factors that can contribute to neurological symptoms. This is something that should be thoroughly discussed with the healthcare team.</p> <p>Typically, neurological symptoms arise when individuals have either exceptionally high levels of phytanic acid or experience a sudden spike, possibly due to recent illness or weight loss. In such cases, the nerves may become highly irritated by this spike. It's advisable to wait for improvement, especially if the levels are beginning to decrease. However, it's essential to recognize that there are individuals who have had Refsum disease for an extended period and continue to experience neurological symptoms. These cases require comprehensive evaluation beyond just dietary considerations.</p>

Question 10	Any advice on the portion size of a carbohydrate snack such as before exercise or bedtime?
Answer	<p>Nutritional information labels on food products often provide the amount of carbohydrate per 100g and per portion. Foods containing carbohydrate can be either starchy foods, like bread, potatoes, rice and pasta, or foods rich in natural sugars like fruit, yoghurt, milk and fruit juice and foods with added sugars such as cakes, biscuits and desserts. For snacks, please consider consuming approximately 10 to 20 grams. For meals, aim for a higher intake, around 70 to 80 grams.</p> <p>Before engaging in exercise, it's recommended to consume 15-20 grams, such as a small carton of fruit juice, which typically contains around 15 grams. Alternatively, a large banana provides approximately 20 grams, while a medium or thick slice of toast also offers about the same amount, around 20 grams.</p>

Question 11	At our center, we see Refsum patients as well as AMACR patients. We advise the same diet. It would be so helpful if the pristanic acid values would also be evaluated if possible.
Answer	Pristanic acid has been analysed as part of the food analysis – please contact Sarah and Eleanor to discuss this further.

Question 12	If someone has "mastered" the Refsum diet, would you recommend continuing to avoid free phytol foods (which, if I understand correctly, are no longer included in either level)
Answer	We recommend following the most recent dietary recommendations as we think it is important for a good quality of life to follow the least restrictive diet possible. This suggests you could try the new dietary guidelines and then review your response.

Question 13	I know that we are now allowed to have multi grain bread, but does it matter which grains are consumed? I am particularly interested in your thoughts on quinoa, buckwheat, and millet?
Answer	We do not know the answer to that question. Currently no restriction of these grains is advised because there is no analytical data to suggest restriction is necessary.

Question 14	I know the new guidelines say we can have all vegetables in level one. But if we are following level 2, what do we do about vegetables that have not necessarily been tested? For example, zucchini and I am also interested about parsley as I love tabouli. I love the fact that the green section of level 1 says we can eat all vegetables, but I still need to be a bit more cautious than that and I am not sure how to go about that with this new diet. Before I knew I had Refsum I used to eat so much kale and then I was so upset to find out how bad it is for me and I just don't want to make that same mistake and I don't know how cautious to be.
Answer	We recommend you continue to eat the vegetables you like until further research suggests that restriction would be beneficial. If you are needing to follow Level 2 advice, we would recommend only limiting the vegetables listed on the diet guide.

Question 15	Why is coley permitted again while cod and haddock aren't?
Answer	The dietary analysis results show that Cod and Haddock contain more phytanic

Question 16	Please clarify the type of perch fish permitted as there are many different types. For example, pike perch, which is very low in fat, see bass, ocean perch and tilapia. Are all types permitted?
Answer	Only one form was analysed. It has been described as fully as possible.

Question 17	I am interested to know whether almond protein still contains phytanic acid, as most of the fat has been removed. Many vegan products now only contain almond protein.
Answer	We do not have a definitive answer to this question. However, we do not currently recommend restricting these products.

Question 18	What is the recommended PA level to start gradual weight loss? If levels remain around 500 -700, can we still gradually lose weight (or is it not recommended). If not recommended what are the alternatives?
Answer	We do not recommend weight loss unless your phytanic acid reading is below 300. Any discussions about weight loss need to be with your managing physician as other factors relating to your individual health may need to be taken into consideration. Weight loss will need to be gradual and with close phytanic acid monitoring.

Question 19	If trying to avoid glucose spikes, what is a recommended snack if out walking or generally out?
Answer	Exercise reduces blood glucose spikes. Fresh fruit combined with a protein source such as seeds or nuts is a good low glycaemic index choice which can give flatter blood glucose response curves.

Question 20	If one doesn't have a dietician, what should we pay attention to in order to ensure we consume sufficient protein and carbohydrates, especially when aiming for good phytanic acid values and a slight weight loss? For instance, how do we determine if we only need one slice of bread instead of two?
Answer	Most people need to eat not less than fifty grams of carbohydrate at each meal which would be at least two slices of bread. Nutrition information labels and apps like My fitness pal or Nutracheck can be helpful in providing carbohydrate guidance on the carbohydrate content of food required in relation to energy requirements. A protein rich food is recommended at each meal e.g. eggs, chicken etc.

Question 21	Regarding the emergency plan, can protein shakes be used for that purpose? I like to have them on hand when there's nothing edible at hand. They provide an extra 300 calories per pack! And they're vegan.
Answer	Protein shakes are not suitable for the emergency regimen. In this circumstance, you need to focus on carbohydrates, not protein, to stop phytanic acid release from your stores.

Question 22	It would be ideal if palm oil and rapeseed oil were tested, as these two fats are present in many vegan products, and we are unsure if they are allowed. In the caution list, sausages and salami are included, which both normally contain beef (in Germany), plus sausages are in sheep casings, which are both not allowed.
Answer	Rapeseed oil (canola oil) has been tested and is ok to use. Palm oil has not been tested yet and we hope to test this in the future. That is useful information about the sausages – we will update the diet sheet to specify pork sausages and salami, as ones containing beef will need to be avoided.

Question 23	Crab appears both in the caution and red section in Level 1 guidance. Which one is correct? Or if they are different, what is the difference?
Answer	Thank you for alerting us. The analysis reports were for white meat only or red and white meat. On reflection, as these are difficult to separate, crab should be in the avoid section, we will update the diet sheet accordingly.

Question 24	It seems that pork meat and fish were not as phytanic acid free as previously thought. It was mentioned at the recent webinar that it also depends on what pigs and fish are fed. Can you please provide more information about that, please? E.g. is meat from pigs fed acorns, such as Iberico pigs, safer than that from others?
Answer	We do not know the answer to this question as pig meat has not been analysed by feedstuff. We recommend choosing lean cuts of pork only.

Question 25	Are haricot beans safe to eat or is it the sauce used in particular brands of baked beans that is a problem?
Answer	We do not know the answer to that question as they have not been tested separately.

Question 26	Are corn/maize and corn oil now safe to eat as they are no longer listed?
Answer	Corn oil is on the green list for Level 1 and Level 2 diets.

Question 27	Are products containing buttermilk safe to eat as in UK it is fat free but partly fermented? (just in case the US has a different definition of buttermilk).
Answer	We do not recommend buttermilk. The UK version has not been tested so the only values we can refer to are US values which are high in phytanic acid.

Question 28	When a branded, shop bought food item is included on the diet sheet should that not be referenced so we can check for alternative brands? e.g rice crispies - any cereal brands checked (in the UK) do not list any ingredients on the red or orange list so Kellogg's Rice Krispies are ok
Answer	Very few breakfast cereals have been tested. We have tried to avoid brand information because it makes the guide very difficult to read. Rice Krispies were tested some years ago. We believe they were Kelloggs.