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Current Dietary Practice in the Management of Adults with Familial Chylomicronaemia Syndrome

A UK Expert Panel Opinion Piece

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Introduction

Familial chylomicronaemia syndrome (FCS) is a rare autosomal recessive disorder with an estimated prevalence of 1–2 per million, which equates to 55–110 people in England.[1,2] FCS is characterised by severe fasting hypertriglyceridaemia (HTG), defined as triglyceride (TG) concentrations >10 mmol/L (885 mg/dL) by the European Atherosclerosis Society Consensus Panel, and a build-up of chylomicrons.[2,3] Other symptoms include episodes of abdominal pain, recurrent acute pancreatitis, eruptive xanthomata and hepatosplenomegaly.[1] Loss-of-function mutations in the gene encoding lipoprotein lipase (LPL) underlie FCS in >90% of patients; however, mutations in several other genes encoding associated proteins responsible for proper LPL function, including *APOC2, GPIHBP1, APOA5* and *LMF1*, may also be responsible.[4]

Occurring in up to 42% of patients, acute pancreatitis is one of the most common and potentially severe complications of FCS.[5] Recurrent episodes can result in chronic pancreatitis and exocrine and endocrine functional impairment, including diabetes secondary to pancreatitis, and can be fatal.[6-8]

Dietary management is the mainstay of treatment and requires close monitoring to ensure it is nutritionally adequate. Furthermore, adhering to strict dietary requirements to manage FCS may have psychosocial implications that can influence eating patterns.[9] Dietary management of FCS aims to maintain TG concentrations below 10 mmol/L (885 mg/dL), the level thought to minimise the risk of pancreatitis.[1,3] This is achieved by restricting dietary fat to minimise chylomicron production and may require complementary interventions, such as TG-lowering therapy.[1] Additionally, patients in the UK with genetically confirmed FCS who are at high risk of pancreatitis and do not respond to dietary management or TG-lowering therapy may be eligible for treatment with an antisense oligonucleotide as an adjunct to dietary management.[2,10]

To enable dietitians to provide appropriate care and support for patients with FCS, clear dietary recommendations are required that consider the changing needs of the patient as they transition from childhood to adulthood and beyond. Until 2018, the only published guidelines available for managing patients with FCS focused on infants, children and adolescents and there was a gap in the literature for best-practice guidelines that addressed the needs of patients with FCS throughout their lifetime.[11] A panel of US-based registered dietitian nutritionists (RDNs) published a series of recommendations based on their collective experience of managing these patients and the scientific literature available.[9] These guidelines provide a basis for dietary management of patients with FCS; however, their application to the UK setting has not been reviewed. To address this limitation, a group of UK-based dietitians convened to review these guidelines and, through the process of a survey, attendance at a series of meetings and email correspondence, they reached a consensus on how to adapt and adopt them in the UK setting. Recommendations based on the current practice of UK-based dietitians for managing adults with FCS, including additional considerations for special populations, are outlined below. Further research is needed to inform the next steps for developing evidence-based consensus dietary guidelines for the management of patients with FCS. Additional dietary support resources for patients and recipe suggestions are available via the UK-based patient support group Action FCS (www.actionfcs.org).

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Dietary recommendations

Core recommendation
These five core recommendations are applicable to all adult patients with FCS.

Core recommendation 1

- Adopt a minimal-fat diet by restricting total dietary fat (excluding medium-chain triglyceride [MCT] oil/powder) to 10–15% of total daily required energy intake
  - This recommendation should be used in addition to regular (preferably fasting) TG concentration monitoring and may require further adaption depending on TG levels.

Rationale for this recommendation

The purpose of the minimal-fat diet is to reduce TG levels below 10 mmol/L (885 mg/dL), although below 20 mmol/L (1,770 mg/dL) is acceptable if no symptoms are present, to minimise the risk of acute pancreatitis episodes. In the absence of a known TG concentration, a daily fat allowance of 10–15% of the total required caloric intake would be a good starting point for most patients; however, recommendations regarding daily fat intake should be guided by each individual patient's TG concentration. This is important as both the disease severity and the amount of fat that can be tolerated is likely to vary between patients. A daily fat allowance as a percentage of the total caloric intake has been recommended (as opposed to a set amount of fat in grams per day as utilised by Williams et al) to facilitate individualisation based on the dietary needs of each patient.

Core recommendation 2

- Aim to meet minimum essential fatty acid (EFA) requirements of 2.5% of the total daily energy intake derived from omega-6 polyunsaturated fatty acids (PUFAs), specifically linoleic acid, and 0.5% from omega-3 PUFAs, specifically alpha linolenic acid, in the minimal-fat diet.

Rationale for this recommendation

EFAs cannot be produced by the body and must be obtained through the diet to prevent deficiency. The recommended intake is 10–16 g/day and 1–3 g/day (depending on age and gender) for omega-6 and omega-3 fatty acids, respectively,[12, 13] which is a large proportion of the daily fat allowance for a patient following a minimal-fat diet. Consequently, the recommendation should be set to target the lowest possible amount necessary to meet EFA requirements. Core recommendation 2 is aligned with the World Health Organization (WHO) recommendation for the minimum EFA consumption to prevent symptoms of deficiency.[14] If the recommendation of 2.5% of the total daily energy intake derived from omega-6 PUFAs is achieved, then supplementation of arachidonic acid is unlikely to be required.

Conversion of omega-3 to docosahexaenoic acid (DHA) and eicosapentaenoic acid (EPA) is varied and often poor. If patients are consuming the minimum amount of omega-6 and omega-3 per day, then it might be necessary to supplement with pre-formed DHA and EPA at 250 mg/day as per WHO recommendations. Dietary sources of EFA include chicken breast, leafy green vegetables, wholegrains, and fortified foods; however, if a patient is unable to meet EFA requirements through dietary sources, clinical practice has shown that a small dose of walnut oil can ensure that these requirements are met. Dietary analysis can be helpful to monitor dietary intake of EFAs. If available, regular blood monitoring of EFAs is important to inform dietary and supplementation requirements.

Evidence for the benefit of high-dose omega-3 supplementation for lowering TG levels in patients with FCS is inconsistent;[4, 15-17] however, if high-dose omega-3 supplements are included as part of the medical management, this should be considered when determining the daily dietary omega-3 intake.

Core recommendation 3

- Prescription-grade MCT oil/powder may be used as a source of energy in the diet and to allow for adjustment of energy intake and macronutrient composition of the diet.

Rationale for this recommendation

MCTs are metabolised through a chylomicron-independent pathway;[18] therefore, MCT products can be used in the dietary management of patients with FCS to increase the overall caloric intake and prevent excessive caloric intake from carbohydrate sources. Only prescription-grade MCT products are recommended for use; over-the-counter products and/or coconut oil should be avoided because these products may also contain long-chain triglycerides (LCT) and therefore may not be a pure source of MCTs.

MCT products should be incorporated into the diet gradually. Close monitoring of the patient is required to assess any side effects, such as gastrointestinal disturbances. Support and guidance in relation to how to incorporate MCT products into the diet is recommended.

Core recommendation 4

- Take a daily multivitamin supplement to provide reference nutrient intake (RNI) for vitamins A, D, E and K with the meal with the highest fat content to optimise absorption. Monitor vitamin levels by biochemical analysis on an annual basis, or more frequently if deficiency is suspected.

Rationale for this recommendation

A fat-restricted diet may limit provision and uptake of fat-soluble vitamins; therefore, supplementation of vitamins A, D, E and K may be required to avoid deficiency. If biochemical monitoring indicates deficiency of any fat-soluble vitamin(s), this should be replaced and the patient monitored more frequently.

Core recommendation 5

- Abstain from consuming alcohol.

Rationale for this recommendation

Avoidance of alcohol is necessary to prevent acute elevations in TG concentration and exacerbation of acute pancreatitis episodes. Non-alcoholic soft drinks and alcohol-free beers and spirits that are low in sugar could be recommended as alternative options.

Secondary recommendations

If the adult patient is successfully managing to follow the minimal-fat diet as outlined in the core recommendations; the following secondary recommendations may be considered.

Secondary recommendation 1

- Aim for up to 25% of the total daily energy intake from protein to be derived from low-fat protein sources.

Rationale for this recommendation

In addition to meeting protein requirements, ensuring adequate protein is important to support satiety on a minimal-fat diet. Protein-rich foods can contain fats and therefore providing information/resources on lean protein options is important to ensure adults with FCS can meet their protein requirements.

"Due to the restrictive nature of the diet to manage FCS during pregnancy, supplementation with a multivitamin that provides at least 400 µg folic acid and 10 µg vitamin D is recommended"

Secondary recommendation 2

- Limit simple and refined carbohydrate foods.

Rationale for this recommendation

The primary focus of the minimal-fat diet is to limit fat intake to reduce TG levels to below 10 mmol/L (885 mg/dL). However, patients following a minimal-fat diet tend to consume a large quantity of refined carbohydrates because these foods often have a lower fat content compared with complex carbohydrate sources. Consequently, patients may consume a large amount of sugar, which can also lead to an increase in the TG concentration. Providing guidance on portion sizes of carbohydrates can be useful to reduce the impact of high carbohydrate intake on TG levels. Examples of complex carbohydrate foods include oats, wholegrain and wholemeal varieties of bread and cereals, and brown rice.

Recommendations for specific life stages/circumstances

Additional dietary advice/guidance is likely to be needed for patients with FCS at particular life stages or in specific circumstances. In all cases, it is necessary to ensure that the diet is tailored to the individual patient and their cultural/religious/dietary needs. Currently, limited dietary resources are available for patients with FCS and further development of more diverse resources is required to suit all patient groups. However, the following recommendations are applicable to adult patients with FCS during pregnancy, those who have concurrent diabetes and/or pancreatic enzyme insufficiency, and those with chronic pancreatitis.

Patients with FCS during pregnancy

Patients with FCS during pregnancy: recommendation 1

- Adopt a minimal-fat diet by restricting total dietary fat (excluding MCT oil/powder); the daily fat intake should be individualised and may need to be adjusted throughout the different trimesters of pregnancy. Consequently, regular monitoring of TG levels is essential throughout pregnancy to ensure appropriate dietary management.

Rationale for this recommendation

In the general population, TG levels are shown to increase during pregnancy; the TG concentration may increase 2- to 4-fold by the third trimester and this is further exacerbated in pregnant women with FCS.[19] Case reports of pregnancies in patients with FCS have shown that fat intake may need to be restricted to 1–2% of the caloric requirements in the later trimesters along with initiating pharmacological therapies, such as fibrates, to manage TG levels and reduce the risk of acute pancreatitis.[20-27] MCT oil/powder and fat-free oral nutritional supplements will likely be required to ensure that adequate calories are obtained while following this minimal-fat diet.

Patients with FCS during pregnancy: recommendation 2

- Ensure EPA and DHA requirements are met; supplement with 300 mg of EPA and DHA per day, of which at least 200 mg should be DHA.

Rationale for this recommendation

Pregnant and lactating females require higher levels of EFAs than stipulated in the core recommendation for adults with FCS to support foetal and infant development. This can be achieved with an over-the-counter omega-3 supplement that is appropriate for use during pregnancy (i.e., does not contain fish liver oil). The above is aligned with WHO recommendations for EFAs during pregnancy and lactation.[12] If it is necessary to restrict fat intake to <10% of the daily caloric requirement, meeting the omega-6 and omega-3 requirements orally during pregnancy will be challenging. Topical application of sunflower oil has been shown to provide a source of these EFAs.[25] If available, regular blood monitoring of EFAs is important to inform dietary and supplemental requirements throughout pregnancy.

Patients with FCS during pregnancy: recommendation 3

- Ensure adequate vitamin and mineral intake for the health of the pregnant woman and normal foetal development.

Rationale for this recommendation

As with all pregnancies, adequate vitamin and mineral intakes are important for normal foetal development. Due to the restrictive nature of the diet to manage FCS during pregnancy, supplementation with a multivitamin that provides at least 400 µg folic acid and 10 µg vitamin D is recommended. With regard to fat-soluble vitamins, non-pregnant women with FCS are advised to meet the RNI for vitamin A, D, E and K prior to conceiving. When planning a pregnancy and throughout pregnancy, plasma fat-soluble vitamin levels should be monitored regularly and supplementation adjusted to ensure levels stay within the normal reference range. Caution is required regarding vitamin A intake during pregnancy.

Patients with FCS during pregnancy: recommendation 4

- Increased multidisciplinary team (MDT) support is recommended for pregnant women with FCS and underlying diabetes or who have developed gestational diabetes.

Rationale for this recommendation

Women with FCS and underlying or gestational diabetes will require additional MDT support during pregnancy due to the increased challenges of managing TG levels during this time. Early screening for gestational diabetes would be beneficial to support early dietary interventions. Dietary recommendations for diabetes management in FCS are covered below.

Patients with FCS during pregnancy: recommendation 5

- Ensure adequate calories are consumed and vitamin and mineral requirements are met to support lactation postpartum.

Rationale for this recommendation

It is important to ensure that women who wish to breastfeed receive adequate calories to support milk production. MCT oil/powder and fat-free nutritional supplements may be beneficial to ensure adequate calories are consumed while adhering to the minimal-fat diet. Women with FCS who are breastfeeding should meet the same vitamin and mineral requirements as the general population for lactation. As per core recommendation 4, women should take a daily multivitamin supplement to provide RNI for vitamins A, D, E and K. Supplementation with EPA and DHA, as per core recommendation 2, should continue during lactation. Regular monitoring of weight, intake and nutritional bloods (including fat-soluble vitamins) is recommended and supplementation should be adjusted as needed.

Patients with FCS and concurrent diabetes and/or pancreatic enzyme insufficiency

Patients with FCS and concurrent diabetes and/or pancreatic enzyme insufficiency: recommendation 1

- Ensure that a multivitamin and mineral supplement is taken with the meal with the highest fat content and that adequate pancreatic enzyme replacement is provided. The advice of a specialist practitioner/dietitian with expertise in pancreatic enzyme replacement therapy should be sought for dosing recommendations and monitoring of tolerance.

Rationale for this recommendation

The risk of fat-soluble vitamin deficiency in patients with FCS who are following a minimal-fat diet is further heightened in patients with concurrent malabsorption through reduced pancreatic exocrine function. The above recommendation is intended to mitigate this risk. It would also be advisable to adopt the same approach to EFA supplementation.

Patients with FCS and concurrent diabetes and/or pancreatic enzyme insufficiency: recommendation 2

- If a patient is using MCT oil/powder to increase daily caloric intake and prevent excessive caloric intake from carbohydrate sources, seek input from a dietitian who has expertise in pancreatic enzyme replacement therapy if the patient shows signs of malabsorption. Signs of malabsorption may include bloating, wind, steatorrhoea and weight loss.

Rationale for this recommendation

There is inconclusive evidence regarding the role of pancreatic enzymes for the absorption of MCTs.[28] MCT digestion bypasses the usual fat digestion process so is not dependent on pancreatic enzyme activity; however, some studies suggest pancreatic enzyme therapy may help to optimise absorption of MCTs. Some over the counter MCT products are not pure and may contain LCTs; therefore, it is recommended that a pure form of MCT is prescribed.

Patients with FCS and concurrent diabetes and/or pancreatic enzyme insufficiency: recommendation 3

- Encourage low glycaemic index (GI) foods at meals and snack times.

Rationale for this recommendation

Simple and refined carbohydrate foods can lead to glycaemic instability and result in hyperglycaemia. The GI of a food can be lowered by the addition of protein and/or MCTs. In patients with concurrent diabetes the glycaemic load of a meal (a measure of the amount of carbohydrate in a portion of food together with how quickly it raises blood glucose levels) should also be considered. It may be beneficial to limit portion sizes of carbohydrate foods to support blood glucose management. Practise caution when referring to GI lists, as low GI foods may be high in fat so it is important to review low GI suggestions in the context of the minimal-fat diet.

"Ensure that a multivitamin and mineral supplement is taken with the meal with the highest fat content and that adequate pancreatic enzyme replacement is provided"

"For patients with FCS experiencing an acute episode of pancreatitis, clinicians should implement further fat restrictions on a short-term basis depending on severity of disease, biochemical, clinical and anthropometric indicators."

Patients with FCS and concurrent diabetes and/or pancreatic enzyme insufficiency: recommendation 4

- Aim for optimal control of blood glucose levels through titration of relevant medications with input from a specialist diabetes centre.

Rationale for this recommendation

Dietary recommendations for patients with concurrent diabetes (regardless of cause) will vary depending on the type of diabetes and treatment. Poorly controlled diabetes can in turn lead to elevated TG levels in patients with any type of diabetes; therefore, it is important for patients following a minimal-fat diet whose TG levels remain elevated that their diabetes control is also evaluated. There may be a lower threshold for commencing insulin therapy in patients with concurrent diabetes and raised TG concentrations. Metformin should be prescribed to reduce blood glucose levels and may have a secondary effect by lowering TG levels. If a patient has diabetes secondary to pancreatitis and blood glucose levels vary markedly, the patient should be assessed for pancreatic enzyme insufficiency. If the patient is already receiving pancreatic enzyme replacement therapy, then dosing should be reviewed.

Patients with FCS and chronic pancreatitis

Patients with FCS and chronic pancreatitis: recommendation 1

- Further reduce daily fat intake until symptoms are under control and enzyme replacement therapy regimen has been established (if required).

Rationale for this recommendation

In contrast to the European Society for Clinical Nutrition and Metabolism (ESPEN) guideline, which states that fat restriction in patients with chronic pancreatitis is only indicated if they have steatorrhoea symptoms that cannot be controlled,[29] patients with FCS and chronic pancreatitis may require further fat restrictions. Such restrictions should be individualised dependent on biochemical markers and severity of disease. As stated in the previous section, if a patient has also been diagnosed with pancreatic enzyme insufficiency, pancreatic enzyme replacement therapy should be initiated following thorough nutritional assessment and titrated to the individual and their daily fat intake. The advice of a specialist practitioner/dietitian with expertise in pancreatic enzyme replacement therapy should be sought for dosing recommendations and monitoring of tolerance.

For patients with FCS experiencing an acute episode of pancreatitis, clinicians should implement further fat restrictions on a short-term basis depending on severity of disease, biochemical, clinical and anthropometric indicators. The duration of these further fat restrictions is subject to patient response and should be implemented on an individual basis.

Patients with FCS and chronic pancreatitis: recommendation 2

- Increase the frequency of monitoring vitamin levels by biochemical analysis as patients with FCS and chronic pancreatitis may have a greater risk of deficiency; consider increasing the dose/frequency of vitamin supplementation.

Rationale for this recommendation

Standard nutritional assessment, including screening for deficiency of fat-soluble vitamins (A, D, E and K), trace elements (magnesium, selenium, zinc), anaemia (iron studies, B12, folate, ferritin), glycated haemoglobin (HbA1c), and random blood glucose should be considered at the diagnosis of chronic pancreatitis.[29] More frequent monitoring may be required if deficiency is suspected.[29] If chronic deficiency of fat-soluble vitamins is observed, supplementation should be initiated with dosing and monitoring implemented as clinically indicated. Further guidance regarding supplementation may be obtained from a metabolic clinical specialist.

Patients with FCS and chronic pancreatitis: recommendation 3

- Standard nutritional assessment should be carried out by a dietitian and, if indicated, low-fat oral nutritional supplements should be provided if the patient is losing weight or having difficulty maintaining a healthy weight.

Rationale for this recommendation

Low-fat oral nutritional supplements are likely to be required by patients with FCS following a fat-restricted diet. A peptide-MCT-based supplement may also be appropriate for patients with FCS and chronic pancreatitis and, if this is indicated, pancreatic enzyme replacement therapy should be optimised for absorption as required.

Conclusions

The recommendations in this article have been proposed by an expert group of UK-based dietitians and are intended for use by dietitians involved in the management of UK-based patients with FCS. These recommendations have been informed by the 2018 publication by a US-based expert panel of RDNs intended to provide dietary advice for patients with FCS across the lifespan and reflect the current practices used by dietitians within the UK.[9] However, further research is needed to inform the next steps for developing evidence-based consensus dietary guidelines for the management of patients with FCS within the UK. Diet and lifestyle are key to maintaining TG concentrations below 10 mmol/L (885 mg/dL) and reducing the risk of pancreatitis as well as other symptoms associated with FCS. Even with strict adherence to the minimal-fat diet, some patients may still experience elevated TGs and episodes of acute pancreatitis; therefore, although these recommendations are intended for all patients with FCS (including those at particular life stages or in specific circumstances), it may be necessary to refine or adapt them to suit the needs of the individual patient.

Restricted daily fat intake is the basis of FCS management and may complement medical treatment with an antisense oligonucleotide (for eligible patients) or other therapies intended to manage comorbidities. Although these recommendations are aimed at dietitians, they are intended to form part of a multidisciplinary approach to patient management. It is important that all members of the MDT are aware of the importance of diet for controlling the symptoms of FCS and are able to support patients in adhering to the recommendations outlined in this article. If a patient cannot follow these recommendations and adhere to the minimal-fat diet, it is worth referring them for psychological support to help them cope with their diagnosis and the challenges of managing this lifelong condition.

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