

A Practical Guide for the Dietary Management of Adults with Familial Chylomicronaemia Syndrome (FCS)

FCS is a rare autosomal recessive disorder resulting in severe fasting hypertriglyceridaemia.

Fasting triglycerides (TG) >10mmol/L (885mg/dL)



1-2 per million



Present throughout the lifecycle



The primary and lifelong treatment

Symptoms

- > Recurring acute pancreatitis (the most severe complication) leading to chronic pancreatitis & diabetes
- > Abdominal pain
- > Eruptive xanthomata
- > Hepatosplenomegaly

Treatment Aim

To reduce the risk of acute pancreatitis by **maintaining fasting TG levels below 10mmol/L (885mg/dL).**

Core Recommendations

Secondary Recommendations

Once adherence to all 5 core recommendations is achieved.

1

Very low-fat diet: 10-15% of total energy intake

2

Essential fatty acids: omega-3 and omega-6

3

Use of medium chain triglycerides

4

Fat soluble vitamins: A, D, E & K

5

Alcohol avoidance

1

Low-fat proteins to make up 25% of energy

2

Limit simple and refined carbohydrates

Specialised dietetic input is essential Very low-fat diets can result in nutritional deficiencies. Ideally refer to a lipid specialised centre with access to a dietitian, a metabolic unit dietitian or contact HEART UK.

The Dietary Recommendations

Tailored advice: adapt according to dietary assessments and fasting TG levels

CORE 1: VERY LOW-FAT DIET

10-15% of total energy intake



Reductions in total fat intakes until fasting TG levels are sustained to <10mmol/L (885mg/dL)
e.g., 2,000kcal daily requirement = 22-33g total fat

Important: Distribute fat evenly throughout the day.



- > All fats – from animal and plant sources.
- > Assessment of food diaries and close monitoring of fasting TG levels is essential. Each patient will respond differently to fat reductions.
- > Start at 15% total energy from fat and adapt according to fasting TG levels.
- > The avoidance of acute pancreatitis episodes is a highly motivating factor for adherence.
- > This fat limit does **not** include MCT.

CORE 2: ESSENTIAL FATTY ACIDS

Ensure requirements are met



Essential fatty acids (EFAs) - omega-3 and omega-6 - cannot be made by the body and must be obtained through the diet.

Important: EFA intakes **must be included within the 10-15% energy fat restriction.**

- > Intakes will depend on the level of fat restrictions needed. Those requiring very strict restrictions e.g., 10% energy or less, may require supplementation.
- > If possible, blood levels of EFA should be monitored to assess status.



Which EFA and how much?

- > **Omega-6 PUFA:** a minimum of 2.5% total energy, in particular, Linoleic Acid (LA). This equates to 5.5g per 2,000kcal intake. *EFA intake should be monitored and if an individual's fat restriction allows, a greater EFA intake can be encouraged.*
- > **Omega-3 PUFA:** Alpha Linolenic Acid (ALA) - 0.5% total energy (~1-3g per 2,000kcal).
- > If the patient is struggling to meet intakes, DHA/EPA – long-chain omega-3 supplements of 250mg per day should be considered.

Good sources include:

- > chicken breast, dark green leafy vegetables and wholegrains. *Walnut oil is exceptionally rich in omega-3. ~1 teaspoon walnut oil will help meet requirements.*

Spot deficiency: excessive thirst, frequent urination, rough, dry or scaly skin, dry, dull or 'lifeless' hair and soft or brittle nails. Raised bumps on the skin are particularly characteristic. A patient noticing any of these symptoms will need their diet checked to ensure adequate intakes.

CORE 3: MEDIUM CHAIN TRIGLYCERIDES (MCT)

! MCTs do not impact on serum TG levels, **thus are not included within the fat restriction.**

MCTs are an excellent energy source and indicated when a patient:

- › is struggling to meet energy requirements and/or sustain a healthy body weight, AND/OR
- › needs to reduce carbohydrate intakes (excess carbohydrates can exacerbate TG levels) e.g., if a patient is also diabetic, but calorie intakes need to be sustained.



- › Only **prescription/medical grade MCT oil or powders** should be used. Other commercially available oils including coconut oils will also contain longer-chain fatty acids which will impact on TG levels.
- › Introduce gradually and build up to tolerance.
 - MCT can have gastrointestinal side effects such as bloating, stomach pains, diarrhoea.
- › MCT oils have a very low smoking point and cannot be used in high heat cooking e.g., frying, roasting, baking. Best used after cooking or in raw recipes e.g., dressings.

CORE 4: OPTIMISE INTAKES OF FAT-SOLUBLE VITAMINS A, D, E & K

! Fat soluble vitamins A, D, E & K require fat to be absorbed. **Severe restrictions in fat intake can increase risk of deficiency.**



- › Multivitamin supplements providing A, D, E and K and other vitamins to RNI levels.
 - Not to take individual high dose vitamins – unless medically indicated.
- › To be taken with meals with the highest fat content to optimise absorption.
- › Annual blood level monitoring, especially vitamin D, should be undertaken and supplementation adapted accordingly.

CORE 5: ABSTAIN FROM ALCOHOL

! Alcohol consumption exacerbates TG levels and significantly increases risk of acute pancreatitis.



- › Patients should opt for alcohol-free and sugar-free varieties.

Secondary Recommendations

Once adherence to all 5 core recommendations is achieved.

1: LEAN PROTEINS AT LEAST 25% OF ENERGY

! Increasing protein intakes can help to mitigate the negative impacts of heavily restricting fat in the diet, providing calories and supporting satiety.

MANY PROTEIN FOODS ARE HIGH IN FAT!

Some high fat protein foods that should be avoided include nuts, seeds, egg yolks, oil rich fish and most meat and meat dishes.



Choose low-fat protein foods e.g.:

- › All beans and pulses
- › Quorn (mycoprotein) and soya mince
- › Turkey / chicken breast - skin removed
- › Low fat/lean hams, goat meat
- › White fish, tinned tuna (in brine), prawns
- › Egg whites
- › Fat-free milk, fat-free yogurt, fat-free cheeses such as Quark and fat-free cottage cheese

2: LIMIT SUGARS & REFINED CARBOHYDRATES

! High intakes of refined carbohydrates such as white flour, white bread, white pasta and especially sugars (including honey, all types of syrups, fruit juices) will impact on TG levels.

Important: Carbohydrate should not exceed **60% of total energy intake**. If it does, reduce all carbohydrates.

If this compromises energy intake, increase very low-fat protein foods and consider MCT.



Introduce wholegrains and other high fibre carbs e.g.:

- › Wholemeal pasta, brown rice, wholemeal bread, oats, sweet potatoes and potatoes with their skin

Focus on removing all free sugars

- › Switching to sugar-free beverages including teas and coffees.
- › Keeping fruit juice and smoothies to no more than 150ml per day.
- › Limiting fruit to no more than 2 per day and spreading out through the day.
- › Cut out or minimise all preserves and sugars: all types of sugars, honey, jams/marmalades.
- › Avoid sweets, chocolates, biscuits, cakes, pastries.

Diet Examples

Two days on a 10-15% energy from fat restriction

Example 1	kcal	Fat g
Breakfast <ul style="list-style-type: none"> > Porridge made with 35g oats & 200ml skimmed milk > 1 large banana > Latte with 150ml skimmed milk 	367	4.1
Lunch <ul style="list-style-type: none"> > Baked beans (reduced sugar) on 2 slices wholemeal toast plus 125g mushrooms pan fried in stock (no fat) plus 2 scrambled egg whites > Diet fruit yogurt > Apple 	565	4.2
Dinner <ul style="list-style-type: none"> > A large chicken breast, skin removed, stuffed with fat-free Quark cheese, seasoning and herbs, wrapped in foil, baked in the oven > Broccoli and carrots, steamed or boiled > Boiled new potatoes 	449	4.6
Throughout the day <ul style="list-style-type: none"> > A slice of malt fruit loaf > 1 tsp walnut oil – to top up EFAs > 200ml skimmed milk > 125g pot diet fruit yogurt 	275	6.2
Total	1,656	19 (10.4%)

Plus additional MCT oil to meet calorie needs

Example 2	kcal	Fat g
Breakfast <ul style="list-style-type: none"> > 2 x Weetabix (or similar) with 150ml skimmed milk plus 125g low fat yogurt > Handful blueberries 	300	2.8
Lunch <ul style="list-style-type: none"> > Tuna sandwich: 2 slices wholemeal bread, Branston pickle, 1 can of tuna in spring water (drained), lettuce, cucumber, tomato > Low fat rice pudding 	488	4.8
Dinner <ul style="list-style-type: none"> > Quorn stir fry: 100g Quorn and a mixture of veg cut into strips, fried in a non-stick pan – use stock if things start to stick. > Add a stir fry sauce. > Serve with egg noodles 	479	5.4
Throughout the day <ul style="list-style-type: none"> > 150g Greek-style 0% fat yogurt > 1 small banana > 200ml skimmed milk > 1 tsp walnut oil - to top up EFAs 	261	5.7
Total	1,529	18.8 (11%)

Plus additional MCT oil to meet calorie needs

Frequency of Dietary Consultations

- First appointment (at least an hour)**
 Should be undertaken based on blood results: fasting TG, if possible, essential fatty acids & fat-soluble vitamins.
- Follow-up within 1-2 weeks of the first consultation**
 Face to face or phone/video call.
- Arrange for further appointments as needed (fortnightly, monthly etc.)**
 Practical information e.g., how to read a food label, recipes, eating out etc.
- Once stable, appointments every 6-12 months**

Additional Considerations for FCS at Specific Life Stages or with a Concurrent Disease

Patients with FCS at certain life-stages or with concurrent diseases, will need additional dietary manipulations and significantly tighter monitoring of fasting TGs, EFAs and vitamins to ensure they remain asymptomatic.

- > FCS women who are planning to become or are pregnant or breast feeding
- > FCS with concurrent diabetes +/- pancreatic enzymes insufficiency
- > FCS with concurrent pancreatitis

Specialised dietetic input and a multidisciplinary team approach is critical.

To find out more about FCS, visit the [HEART UK website](https://www.heartuk.org.uk)



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