

Cystic Fibrosis and Gene Modulator Therapy

Nutrition considerations

What is gene modulator therapy?

Cystic Fibrosis (CF) is caused by mutations in the CF gene. These mutations lead to defects in a specific protein called the cystic fibrosis conductance regulator (CFTR) protein. A class of drugs called CFTR modulators target the underlying defect in the CFTR protein to improve the function of the chloride channel. These include Kalydeco, Orkambi, Symdeko and Trikafta.

How to take gene modulators:

- Take approximately **10g of fat** with your gene modulator medication to ensure it is absorbed. See Table 1 for examples of food/drink items that contain this amount of fat.
- When feeling sick/nauseous it is important to continue taking your medications with approximately 10g of fat.
- If you are required to be on a clear fluid diet, 10g of fat can be consumed using 10ml MCT (Medium Chain Triglyceride) oil or 10ml of any other oil (e.g. olive oil, vegetable oil).

Table 1 Examples of food containing 10g Fat

Dose	Food
Morning	<ul style="list-style-type: none"> • ½ tbsp oil, butter or nut butter • 1 tbsp or 1/3 avocado • 1 large glass (300ml) of full cream cow's milk • 2 cups of soy or almond milk • 1 cup (250g) full fat yoghurt • 2 eggs • 1 slice of full fat cheese • ¾ cup of toasted muesli or cereal with seeds & nuts with ½ cup full cream milk • fruit smoothie with 200ml milk +2-3 tbsp yoghurt
Evening	<ul style="list-style-type: none"> • 1 cup of milo or flavoured milk • 15 almonds or cashews • 35 pistachios • 1 small tin of fish • 1 handful (~10) olives • ½ tbsp of any oil (can be used to cook dinner meal, or as a salad dressing)

What to expect when taking gene modulator therapy?

Gene modulator therapy may impact your nutrition status.

You may notice:

- improved appetite
- changes to nutrient absorption
- changes to weight

It is important to stay in touch with your dietitian and medical team to discuss any changes you may be experiencing. For some people gene modulator therapy may require you to make dietary changes. Continue to take all regular CF therapies including enzymes, insulin, vitamins and hydration whilst on gene modulator therapy.

Healthy Eating

Healthy eating is important throughout all life stages. A balanced diet provides vitamins, minerals and nutrients which are essential for the body to function. A CF diet is usually high in fat and energy to prevent underweight and undernutrition, however gene modulator therapy may improve nutritional status and you might need to change your dietary goals. You may be advised to reduce some of the high protein and high energy foods and drinks you once required to maintain your weight. See Table 2 for guidelines for healthy eating and discuss further with your dietitian.

NB* Many healthier foods can be lower in salt content. Ensure you are adding salt to meals and seek assistance if you feel your hydration has changed.

Table 2 Guidelines and tips for healthy eating

Guidelines
1. Try to eat regular meals to avoid overeating <ul style="list-style-type: none">• Choose a meal pattern that works for you – 3 main meals or smaller, more frequent meals.• Try to start the day with breakfast.• Plan your meals to prevent relying on quick, easy options that may not be the healthiest choice.• Try not to skip meals or follow fad diets. This can lead to overeating and unsustainable eating habits. Successful eating habits should be easy to stick to and enjoyable.• Include some form of protein at each meal to prevent hunger between meals e.g. lean meat, tuna, eggs, nuts, cheese or beans.• Learn to recognise your body's signals for hunger and fullness to prevent overeating.• Always ask yourself if you are really hungry or just eating out of boredom.

2. Include plenty of high fibre foods

- Aim to eat 2 serves of fruit and at least 5 serves of vegetables every day.
 - o One serve of fruit is a medium piece of fruit e.g. an apple or 1 cup of canned or chopped fruit.
 - o One serve of vegetables is ½ cup of cooked vegetables, 1 cup of salad or one small potato. Try to limit starchy vegetables like potatoes and sweet potato to one serve each day.
- Choose high fibre breads and cereals instead of low fibre options to keep you feeling fuller for longer e.g. wholemeal/wholegrain products.
- Choose whole fruit instead of juice or dried fruit.
- Fill half your plate with salad or vegetables.
- Vegetables also make great snacks; keep some pre-cut carrot, celery and capsicum sticks in your fridge.

3. Choose healthier fat sources

- All fats are high in energy (also known as kilojoules or calories). Eating high fat foods often leads to eating more energy than you need.
- Use only small amounts of margarine/oil when cooking or preparing foods. Try an olive oil spray or a non-stick fry pan.
- Include healthy fat sources such as nuts, avocado, olives, or fish into your meal choices.
- Limit the intake of unhealthy high fat foods like:
 - o Fatty meats e.g. sausages, chops, bacon
 - o Butter, margarine, lard, oil, ghee
 - o Full cream dairy products
 - o Rich cakes, pastries, pies and chocolate
 - o Fatty processed & fried takeaway
- Include low fat protein foods at each meal to prevent hunger between meals.
- Choose lean meat, chicken & fish – trim the fat from meat & the skin from chicken (before cooking is best).
- Choose low fat & reduced fat dairy products.

4. Reduce sugar intake

- Sugar contains no valuable nutrients but can contribute a lot of extra energy (also known as kilojoules/calories) to the diet.
- Try to reduce your intake of high sugar foods and drinks such as lollies, chocolates, biscuits, cakes, pastries, as well as soft drinks, juice, cordial and flavoured milk.
- Try to cut down on sugar you may be adding to tea, coffee, and breakfast cereals.
- Choose lower sugar varieties of foods e.g. low sugar cereals, reduced sugar yoghurt, plain biscuits.
- Artificial sweeteners and sugar alternatives can provide a safe and low kilojoule option for sweetness.

5. Limit alcohol

- Check with your pharmacist to see if alcohol interacts with any of your medications.
- Alcohol should be used in moderation as it is high in kilojoules/calories and low in nutrients.
- For healthy men and women, it is recommended that you drink no more than two standard drinks on any day.
- A standard drink is a:
 - Schooner (425mL) of light beer
 - Pot (285mL) of heavy beer
 - Small glass (100mL) of wine
 - Nip (30mL) of spirit

6. Portion control

- Fill half your plate with vegetables or salad, one quarter with lean protein and one quarter with carbohydrate-based foods.
- Try to adhere to the below plate guide.
- Serve your meals in the kitchen rather than from shared plates to discourage sneaky seconds.
- Put leftovers into meal-sized portion containers and store in the fridge for later.
- Wait 20 minutes after a meal before you serve yourself more.

Healthy Plate Guide

