Position Statement: Pancreatic enzyme replacement therapy (PERT) shortage – advice for the management of adults with pancreatic exocrine insufficiency

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Position statement and advice for prescribers and patients from the [1]Nutrition Interest Group of the Pancreatic Society of Great Britain and Ireland (NIGPS), [2] Cystic Fibrosis Specialist Group and [3] Gastroenterology Specialist Group, British Dietetic Association.

Endorsed by the British Society of Gastroenterology (Pancreas section); Pancreatic Society of Great Britain and Ireland, Pancreatic Cancer UK, GUTS UK, Cystic Fibrosis Trust, CF Medical Association and the British Dietetic Association.

Issue date: 26th March 2024 (Version 1)

Please ensure you are reading the most up to date version.

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Introduction

The ongoing supply issues surrounding pancreatic enzyme replacement therapy (**PERT** – under the product brands: **Creon®**, **Nutrizym®** and **Pancrex®**) has progressed. These intermittent supply issues mean some people are running out of PERT, or experiencing difficulties or delays in accessing PERT. Therefore, there is a need for clinical and symptom management advice that is different to normal clinical practice. This position paper is designed to meet the needs of the clinicians' managing patients with pancreatic exocrine insufficiency (PEI) and provides advice for prescribers, dietitians and patients.

Pancreatic enzyme replacement therapy is prescribed to support adequate digestion in patients with PEI, most commonly due to pancreatic cancer, pancreatitis and cystic fibrosis (CF). There are many other clinical situations where patients may have primary or secondary PEI, such as type 3c diabetes or following gastrectomy or gastric bypass surgery (1). Regardless of aetiology, the impact of maldigestion varies from person to person in both the type of symptoms and their severity.

Symptoms of untreated PEI may include bloating, excess wind, diarrhoea, crampy abdominal pain, faecal urgency, steatorrhoea (pale floating stools), hard to manage blood glucose levels, vitamin and mineral deficiencies, weight loss and malnutrition (1). These symptoms are usually managed with PERT and will recur if patients are unable to take adequate doses.

There are many clinical impacts of inadequate PERT, which will affect all patients, and the advice in this document is targeted at all patient groups. However, particular care should be taken in ensuring absorption is adequate in patients with:

- Cystic fibrosis on CFTR modulators and anti-rejection medication – as malabsorption may impact the efficacy of their treatment.
- Patients who have had a total pancreatectomy
- Patients who have had head of pancreas surgery for diseases in the peri-ampullary region (including but not limited to: pancreatic / duodenal / distal cholangio / ampullary cancers, neuroendocrine tumours and pre-malignant conditions)
- Patients with pancreatic cancer undergoing chemotherapy – as poor performance status due to malabsorption and malnutrition may prevent patients from undergoing potentially curative or life-prolonging treatments.
- Patients with PEI and insulin dependent diabetes
- Any patient with PEI (including pancreatitis, pancreatic cancer, cystic fibrosis, post gastrectomy etc.,) who are struggling to maintain their nutritional status.

The advice in this paper may be updated as we receive further guidance and expand our experience in managing PEI without adequate PERT.

Please note the advice in this document is designed for adults with PEI, specialist advice should be sought for children with PEI.

Patients with cystic fibrosis will be under the care of a specialist centre, and they should contact their specialist team if they have any concerns.

Advice for Prescribers

Patients with PEI prescribed PERT are likely to need a change in their repeat prescriptions due to the changing availability of products. Unfortunately, this may need to be altered on each prescription depending on product availability.

It may be necessary to provide a range of pancreatic enzymes to meet the needs of patients and repeat prescriptions should be flexible to allow for this. Pharmacists are unable to dispense anything other than the medication prescribed, so patients do have to return to their primary care team to obtain a new prescription when they are unable to access the product prescribed.

The Department of Health and Social Care (DHSC) medicines supply notification (issued 16/2/24 MSN/2024/022) has advised that there should be sufficient stock of Creon 10,000 to cover the deficit in Creon 25,000 and Nutrizym 22 (4)

Table 1: Conversion charts (2)

Creon® 25,000 Dose	Equivalent in Nutrizym® 22	Equivalent in Creon® 10,000	Equivalent in Pancrex® 340mg (8,000 units lipase)	Equivalent in Pancrex® 125mg (2,950 units lipase)	Equivalent in Creon® Micro*	Pancrex® V powder*
1 x Creon 25,000	1 x Nutrizym 22	3 x Creon 10,000	3 x Pancrex 8,000	8 x Pancrex 2,950	5 scoops Creon Micro	½ x 2.5ml spoon
2 x Creon 25,000	2 x Nutrizym 22	5 x Creon 10,000	6 x Pancrex 8,000	16 x Pancrex 2,950	10 scoops Creon Micro	1 x 2.5ml spoon
3 x Creon 25,000	3 x Nutrizym 22	8 x Creon 10,000	9 x Pancrex 8,000	24 x Pancrex 2,950	15 scoops Creon Micro	1½ x 2.5ml spoon
4 x Creon 25,000	4 x Nutrizym 22	10 x Creon 10,000	12 x Pancrex 8,000	32 x Pancrex 2,950	20 scoops Creon Micro	2 x 2.5ml spoon
5 x Creon 25,000	5 x Nutrizym 22	13 x Creon 10,000	15 x Pancrex 8,000	40 x Pancrex 2,950	25 scoops Creon Micro	2 ½ x 2.5ml spoon
6 x Creon 25,000	6 x Nutrizym 22	15 x Creon 10,000	18 x Pancrex 8,000	48 x Pancrex 2,950	30 scoops Creon Micro	3 x 2.5ml spoon

**Mix with a mildly acidic puree (fruit yoghurt / apple sauce), rinse mouth with water and ensure thorough mouth care as at risk of ulceration if powder / granules get stuck in the gums / under dentures*

It may be more practical to combine medications – so for instance if the prescription is Creon® 25,000 x 3 with meals and 2 with snacks.

Taking the PERT throughout the meal rather than all at the start/ middle/ end improves efficacy.

Alternative prescriptions to minimise disruption and pill burden to the patient, and reduce usage of Creon® 25,000 could be:

Creon® 25,000 x 3 with meals and Creon® 10,000 x 5 with snacks

or

Creon® 10,000 x 8 with meals and Creon® 10,000 x 5 with snacks.

or

Creon® 10,000 x 8 with meals, Pancrex® 340mg x 6 with snacks.

or

Nutrizym® 22 x 3 with meals, Pancrex® 340mg x 6 with snacks

or

Pancrex® 340mg x 10 with a meal, Pancrex® 125mg x 16 with a snack

Other clinical management suggestions

- Consider prescribing a **proton pump inhibitor** or **H2 receptor antagonist** to reduce acid degradation of the PERT and optimise efficacy (1).
- In patients who are experiencing loose bowel motions or faecal urgency due to lack of PERT, and **WHO DO NOT HAVE CYSTIC FIBROSIS and where an infective, inflammatory (underlying inflammatory bowel disease) or obstructive cause has been ruled out** - please consider prescribing **loperamide** at a starting dose of 2mg in the morning and working up to 2mg before meals (TDS) if needed. Higher doses may be needed and should be assessed individually. This will slow the gut transit time down and help alleviate symptoms but will not treat malabsorption.
- Distal intestinal obstruction syndrome is a unique feature of CF and is characterised by the accumulation of viscid mucofaecal material in the terminal ileum and caecum. Being a common complication in people with CF, it needs to be considered if someone with CF presents with symptoms following a change in their PERT prescription.
- Be aware that patients on insulin or oral hypoglycaemic agents that can cause hypoglycaemia may experience worsening control and be more susceptible to hypoglycaemia. Regular blood glucose monitoring is helpful, and patients on continuous glucose monitoring should be encouraged to ensure their hypoglycaemic alarm is set.
- If your patient has a gastric feeding tube [Percutaneous Endoscopic Gastrostomy (PEG), Radiologically inserted gastrostomy (RIG) or naso-gastric (NG) tube] then they can receive their PERT through the feeding tube in the form on **Pancrex® V powder**. This should be made up each time, the powder mixed with 20mls of water and flushed down the tube. 2g is 50,000 lipase units, this fills the 2.5ml end of a measuring spoon. This should be flushed through the tube during the meal (1) (Appendix 2). Please note, this dose may need to be escalated.
- In patients who have ongoing bowel symptoms and were undergoing dose escalation to manage this, please only continue to increase the dose until the patient is able to maintain their weight. If they have ongoing bowel symptoms after this, we suggest using **loperamide** / **Buscopan®** and other appropriate medications to manage their symptoms rather than continuing to increase the doses to preserve supplies. **Imodium melts®** may be more effective than loperamide capsules. (Please do not do this for patients with Cystic Fibrosis – advice should be sort from their specialist team)
- Consider that patients with PEI, may also have bile acid malabsorption, small intestinal bacterial overgrowth, coeliac disease etc., please investigate and treat in line with current guidelines (2).
- If patients are unable to access any PERT and are losing weight or have intractable or unmanageable abdominal symptoms, we suggest reducing their oral intake of food significantly and prescribing peptide based oral nutritional supplements instead (**Vital 1.5kcal®** or **Peptisip Energy HP®**). Please note this should be used for a short period until further supplies of PERT can be obtained. We recommend you only prescribe one-week supply at a time, due to the cost of these products. Please note standard oral nutritional supplements are **NOT** suitable without PERT (i.e., **Altraplen® Amyes®**, **Ensure®**, **Foodlink® Fortisip®**, **Fresubin®**). Table 2 shows prescription doses by body weight.
- Consider prescribing a calcium and vitamin D supplement if patients are not already taking one (please seek advice from specialist teams for patients with cystic fibrosis).
- Please be aware that malabsorptive diarrhoea may impact the absorption of other medication.

- Please be aware that Vitamin K is a fat-soluble vitamin and uptake maybe impaired with inadequate PERT – additional monitoring may be needed for patients on anti-coagulation.

Table 2: How many nutritional supplement drinks should be prescribed for patients not able to absorb food and without access to PERT.

Body weight	**Supplements needed per day (Vital 1.5kcal® or Peptisip Energy HP®**
Below 40kg	Contact a dietitian
40 - 50kg	4 x 200ml bottles = 1200kcal
50 - 60kg	5 x 200ml bottles = 1500kcal
60 - 70kg	6 x 200ml bottles = 1800kcal
70 - 80kg	7 x 200ml bottles = 2100kcal
80 - 90kg	8 x 200ml bottles = 2400kcal
Over 90kg	Contact a dietitian

This may under-estimate energy needs, however it should be sufficient for short term nutritional support. If needed for longer and the patient is rapidly losing weight or is very active, add in one more bottle per day. If they gain weight and were not intending to – reduce by 1 bottle per day.

IMPORTANT

Whilst the supply issues are ongoing, please do **NOT** encourage patients to stockpile these medicines, as this will further drive the shortage.

We suggest patients place their prescription requests 2 weeks earlier than usual to give the community pharmacist time to source the medication if available.

If you are issuing multiple prescriptions so patients can try and source different products, please advise them to try to use one pharmacy if leaving prescriptions on a "back order" to try to reduce the risk of large volumes being dispensed by different pharmacies.

In line with the Medicines Supply Notification from the Department of Health and Social Care (issued 16/2/24 MSN/2024/022). Please ensure that only one-month supply is issued at a time, and new patients are started on Creon® 10,000. (4)

If you are hospital based, please liaise with your pharmacy team regarding stock levels and consider issuing "rescue prescriptions" from the hospital **only in the event that high priority patients have run out completely**. We recommend no more than 1-2 weeks supply is issued within a rescue prescription. Larger prescriptions risk depleting hospital supplies.

Sources of further advice

For patients with CF please contact the patients' CF Specialist team. For all other patients please contact your local tertiary hepato-pancreatico-biliary (HPB)/ pancreatic unit. Depending on the local service available, this may be either the specialist HPB/ pancreatic dietitian or pancreatologist. The Specialist Pharmacy Service Medicines Supply Tool can be accessed by Prescribers for more information on supply at: https://www.sps.nhs.uk

Advice for Dietitians

In addition to the above advice, please consider the following:

- Remind patients to spread the dose of their PERT out throughout their meals to optimise absorption – taking some PERT at the beginning, some in the middle and some towards the end of their meal.
- Remind patients to store their enzymes properly – all products should be stored below 25 degrees and some require refrigeration. Excess heat causes irreversible denaturation.
- Only dose escalate PERT in nutritionally compromised patients – liaise with medical teams to access loperamide for those who are weight stable to minimise any diarrhoea (Not appropriate for patients with CF). Ensure obstructive / infectious causes have been ruled out first. Refer patients not responding to treatment back to their managing physician to ensure other causes of diarrhoea have been excluded.
- Use peptide based oral nutritional supplements (ONS). i.e., **VITAL 1.5 kcal®** / **Peptisip Energy HP®** in place of polymeric supplements to reduce the need for PERT with polymeric ONS. **Peptamen® Vanilla** contains less energy (1kcal/ml) **Survimed 1.5kcal OPD®** is currently out of stock and has a lower MCT content, but these could be used if supplies of **VITAL 1.5 kcal®** / **Peptisip Energy HP®** are limited.
- Consider fat free ONS sipped slowly without PERT if the patient is weight stable. If the patient experiences significant abdominal symptoms or weight loss despite this, please swap to peptide based ONS. PERT may still be required for polysaccharide and protein digestion. Fat free ONS are not nutritionally complete and should not be used a sole source of nutrition.
- Use **ProSource Jelly®** / **ProSource Plus®** for additional protein (peptide based).
- Medium chain triglyceride (MCT) lipid products (**Liquigen®** / **MCT oil®**) could be used alongside fat free ONS in patients who need higher energy ONS. Please note that **Elemental 028®** contains 35% MCT and still requires PERT for lipid absorption, but **Emsogen®** can also be considered. This can be concentrated if tolerated.
- Consider 10-20% reduction in dietary fat for patients who are symptomatic.
- Consider reducing dietary fibre if large doses are consumed (>40g fibre per day) as high fibre diets can reduce the efficacy of PERT.
- Ensure patients with diabetes on **insulin** or medications associated with hypoglycaemia (i.e. **Gliclazide**) are regularly monitoring their blood glucose levels and aware of how to treat a hypo.
- Please ensure patients with signs of malabsorption and taking anti-coagulation are highlighted to their managing physician as Vitamin K absorption may be impaired.
- Seek advice from specialist centres for specific advice for patients with CF.
    - CF Dietitians will review and optimise PERT dosing and adherence.
    - CF dietitians will optimise vitamins and minerals and adjust as appropriate.
- For patients without CF - If the steps within this guidance are not adequate, please contact your local tertiary pancreatic centre for further advice.
- Patients with intractable malabsorption may need peptide enteral or in severe cases parenteral nutrition.

- If you work in a tertiary pancreatic centre, please liaise with your pharmacy team regarding stock levels and consider issuing "rescue prescriptions" from the hospital in the event that high priority patients have run out completely. We recommend no more than 1-2 weeks supply issued within a rescue prescription if possible. Larger prescriptions risk depleting hospital supplies.

Nutritional composition of additional products

Liquigen® 30mls = 136kcal, 0g protein, 97.4% MCT. ACBS approved. 250ml bottle, once open store in a refrigerator and use within 14 days. Suitable for vegans and vegetarians.

Nutricia MCT oil® 100mls = 855kcal, 0g protein, 99.9% MCT. ACBS approved. 500ml bottle, once open, reseal and use within 1 month. Suitable for vegans and vegetarians.

Vital 1.5kcal® 300kcal, 13.5g protein (as peptide), 64% MCT. ACBS approved. 200ml bottle, suitable for vegetarians (not vegan), once open store in a refrigerator and discard after 24 hours

Peptisip Energy HP® 300kcal, 15g protein (as peptide), 60% MCT, ACBS approved, 200ml bottle, suitable for vegetarians (not vegan), once open store in a refrigerator and discard after 24 hours.

Emsogen® 438 kcal, 12.5g protein per 100g,(as amino acid), 83% MCT, 88kcal per 100mls, 2.5g protein, ACBS approved. Note this is a low energy, low protein supplement drink. Standard concentration is 20% w/v (1 x 100g sachet in 500mls water). This can be concentrated further if tolerated, but patients may require additional fluids afterwards. Milkshake / coffee syrups can be added to flavour.

ProSource Jelly® 90kcal, 20g protein (as peptide), 0g fat, <1g carbohydrates, ACBS approved. 118g serving, serve chilled.

ProSource 20® 90kcal, 20g protein (as peptide), 0g fat, 2g carbohydrates. ACBS approved. 60ml serving, take straight from cup or add to hot/cold drinks.

ProSource Plus® 100kcal, 15g protein (as peptide), 0g fat, 11g carbohydrates. ACBS approved. 30ml serving, flavoured products can be taken as a shot, unflavoured can be added to drinks or food.

Peptamen Vanilla® 200kcal, 8g protein (as peptide), 68% MCT, ACBS approved. 200ml bottle, best served chilled, once open refrigerate and consume within 24 hours.

Survimed® **OPD 1.5kcal** drink – out of stock at the time of going to print.

Sources of product information

Liquigen (nutricia.co.uk)
MCT Oil (nutricia.co.uk)
Vital 1.5kcal | Patient Samples Abbott Nutrition UK
Peptisip Energy HP (nutricia.co.uk)
Emsogen (nutricia.co.uk)
Nutrinovo - ProSource Jelly nutrition supplement - Nutrinovo
Product: ProSource 20 - Nutrinovo
Nutrinovo - ProSource Plus nutrition supplement - Nutrinovo
Peptamen Vanilla Liquid Sip Feed | Nestlé Health Science (nestlehealthscience.co.uk)
Survimed OPD 1.5kcal Drink Cappuccino | Disease Specific (fresubin.com)

References / sources of further information

1) Phillips ME, Hopper AD, Leeds JS*, et al* Consensus for the management of pancreatic exocrine insufficiency: UK practical guidelines *BMJ Open Gastroenterology* 2021 ;**8:**e000643. doi: 10.1136/bmjgast-2021-000643
2) https://bnf.nice.org.uk/drugs/pancreatin/ accessed 16/3/24
3) https://cks.nice.org.uk/topics/diarrhoea-adults-assessment/ accessed 16/3/24
4) Medicine Supply Notification: Creon 25000 MSN/2024/022 Issued 16/02/24
5) A5_Hypo_TREND.pdf (trenddiabetes.online) accessed 16/3/24

Appendix 1: Advice for patients

Patient information is being produced by Pancreatic Cancer UK, GUTS UK and the Cystic Fibrosis Trust, and will be available shortly. In the meantime, the below document can be used to guide patient consultations, and for patients with other medical conditions.

Position Statement: Pancreatic enzyme replacement therapy (PERT) shortage – advice for adults with pancreatic exocrine insufficiency

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INTRODUCTION

The ongoing supply issues surrounding pancreatic enzyme replacement therapy (**PERT** – under the product brands: **Creon**®, **Nutrizym**® and **Pancrex**®) has progressed. These intermittent supply issues mean some people are running out of PERT, or experiencing difficulties or delays in accessing PERT. This position paper is designed provides advice to minimise the impact on your symptoms and quality of life should you be unable to access your normal supply of PERT.

Pancreatic enzyme replacement therapy is prescribed to support adequate digestion in people with pancreatic exocrine insufficiency (PEI), most commonly due to pancreatic cancer, pancreatitis and cystic fibrosis (CF). There are many other clinical situations where people may have primary or secondary PEI, such as type 3c diabetes or following gastrectomy or gastric bypass surgery (1). Regardless of the cause of the PEI, the types of symptoms and their severity will vary from person to person.

Symptoms of untreated PEI may include bloating, excess wind, diarrhoea, crampy abdominal pain, urgency to open bowels, steatorrhea (pale floating stools), hard to manage blood glucose levels, vitamin and mineral deficiencies, weight loss and malnutrition (1). These symptoms are usually treated by taking PERT and will recur if you are unable to take enough.

The advice in this paper may be updated as we receive further guidance and expand our experience in managing PEI without adequate PERT.

Please note the advice in this document is designed for adults with PEI, specialist advice should be sought for children with PEI.

People with cystic fibrosis will be under the care of a specialist centre, some of the advice in this leaflet is not suitable for people with CF, and this has been highlighted. If you have CF you should contact your specialist team if you have any concerns.

We have divided the advice for patients into 4 phases, depending on the supply available.

- Phase 1 – Supplies available
- Phase 2 – Limited supplies
- Phase 3 – Insufficient supplies and you have abdominal symptoms
- Phase 4 – No supplies available

Pancreatic enzymes are still being delivered regularly into the United Kingdom, so we will move backwards and forwards between these phases. We are not anticipating that any individual patient will run out of PERT completely for very long. So, if we do have to use the advice in phase 3 or 4, we think this will only be for a short period of time.

Phase 1: supplies available

Whilst the supply issues are ongoing, please do not stockpile these medicines, as this will further drive the shortage.

The Department of Health and Social Care has recommended that only 1-month supply is issued at a time to try and regulate supplies, so you if you currently receive 2-3 months of your PERT at a time, you will need to collect your prescriptions more frequently. (If you pay for your prescription consider applying for a pre-payment certificate to reduce the cost of prescription charges – NHS Prescription Prepayment Certificate (PPC) | NHSBSA)

We suggest you place your prescription requests **2 weeks earlier** than usual to give the community pharmacist time to source your medication.

You may need a change in your repeat prescriptions if what you usually have is not available. There are three brands of PERT available currently in the UK.

Remember to store your PERT appropriately. All PERT should be stored below 25 degrees, and some brands recommend refrigeration. If PERT gets too hot it does not work properly, this damage cannot be reversed.

Taking the PERT throughout the meal rather than all at the start/ middle/ end improves how well it digests the food and drinks you are eating / drinking.

Ensure that you use your PERT before it goes out of date. If you store PERT in different places (i.e. at work), make sure you rotate your supplies to prevent any wastage.

This table shows how each product compares to others.

Table 1: Conversion charts (2)

Creon® 25,000 Dose	Equivalent in Nutrizym® 22	Equivalent in Creon® 10,000	Equivalent in Pancrex® 340mg (8,000 units lipase)	Equivalent in Pancrex® 125mg (2,950 units lipase)	Equivalent in Creon® Micro*	Pancrex® V powder*
1 x Creon 25,000	1 x Nutrizym 22	3 x Creon 10,000	3 x Pancrex 8,000	8 x Pancrex 2,950	5 scoops Creon Micro	½ x 2.5ml spoon
2 x Creon 25,000	2 x Nutrizym 22	5 x Creon 10,000	6 x Pancrex 8,000	16 x Pancrex 2,950	10 scoops Creon Micro	1 x 2.5ml spoon
3 x Creon 25,000	3 x Nutrizym 22	8 x Creon 10,000	9 x Pancrex 8,000	24 x Pancrex 2,950	15 scoops Creon Micro	1½ x 2.5ml spoon
4 x Creon 25,000	4 x Nutrizym 22	10 x Creon 10,000	12 x Pancrex 8,000	32 x Pancrex 2,950	20 scoops Creon Micro	2 x 2.5ml spoon
5 x Creon 25,000	5 x Nutrizym 22	13 x Creon 10,000	15 x Pancrex 8,000	40 x Pancrex 2,950	25 scoops Creon Micro	2 ½ x 2.5ml spoon
6 x Creon 25,000	6 x Nutrizym 22	15 x Creon 10,000	18 x Pancrex 8,000	48 x Pancrex 2,950	30 scoops Creon Micro	3 x 2.5ml spoon

Please check the storage recommendations on the label
– Some products may need refrigerating

It may be most practical to combine medications – so for instance if the prescription is Creon® 25,000 x 3 with meals and 2 with snacks, these are all equivalent using different products.

Creon® 25,000 x 3 with meals and Creon® 10,000 x 5 with snacks

or

Creon® 10,000 x 8 with meals and Creon® 10,000 x 5 with snacks.

or

Creon® 10,000 x 8 with meals, Pancrex® 340mg x 6 with snacks.

or

Nutrizym® 22 x 3 with meals, Pancrex® 340mg x 6 with snacks

or

Pancrex® 340mg x 10 with a meal, Pancrex® 125mg x 16 with a snack

Phase 2: limited supplies

First Step if supplies are limited:

- Ensure you are taking a proton pump inhibitor (**omeprazole**® / **pantoprazole**®/ **lansoprazole**®) or an (H_2)-receptor antagonists (**Famotidine**® / **Nizatidine**®) – these reduce the acid in your stomach and make the enzymes more efficient. This means a lower dose than your usual dose may be effective if you have a proton pump inhibitor as well. If this does not appear to be effective, they may be stopped.
- Reduce the dose of PERT with snacks before reducing your doses with meals as meals tend to be more nutritious.
- **Reduce the dose of PERT by one capsule with each meal and snack, rather than skip whole meal doses**.
- If you have some high dose PERT left, take this with you when you go out, and use the low dose capsules when you are at home to reduce the number of capsules you need to take out with you.
- Prioritise meals that have the most protein and energy in them.
- If you are not already taking vitamins and minerals, we recommend that you take a calcium and vitamin D supplement (containing 800iu Vitamin D and at least 500mg Calcium) and a multi-vitamin and mineral during this time (i.e. **Sanatogen A-Z Complete**® / **Centrum Advance**® / **Supermarket own A-Z brand** – please ensure these contain both vitamins and minerals)
    - Patients with CF should remain on their prescribed vitamins and minerals and discuss any concerns with your CF specialist dietitian.
- Please contact your dietitian / nurse specialist or doctor if you are struggling with malabsorption symptoms or are consistently losing weight.

If you have diabetes

Monitor your blood glucose levels regularly: before meals, before bed, if feeling unwell, if you feel like you are having a hypo* or before driving. If you are driving long distances, make sure you check your blood glucose levels regularly.

If you take less enzymes with your food, you are likely to absorb less carbohydrate from it. Therefore, you may need to reduce the amount of quick acting or mixed insulin you inject to prevent a hypo*.

*A hypo is when your blood glucose level goes below 4mmol/l, typical symptoms include sweating, shaking, blurred vision, confusion, palpitations. Always keep hypo treatment on you.

You can find more information on recognising and treating a hypo from your diabetes team or by following this link: A5_Hypo_TREND.pdf (trenddiabetes.online). You are only at risk of a hypo if you take medication that lower your blood glucose levels. It is not usually possible to have a hypo if you have diet-controlled diabetes, or diabetes treated with **metformin/DPP-4 inhibitors (Gliptins)**.

If you have a continuous glucose monitor, ensure you have the hypoglycaemic alarm set. If you are having more hypos than usual, you may need to contact your CF/diabetes team for advice on adjusting your insulin doses.

If you take oral nutritional supplements (i.e., **Altraplen® Amyes®**, **Ensure®**, **Foodlink® Fortisip®**, **Fresubin®**), ask your dietitian if they can be changed to a peptide / semi-elemental preparation (i.e., **Vital 1.5kcal®**, **Peptisip Energy HP®**) as many people can manage these without additional enzymes.

These do not come in a wide range of flavours, but you can add milkshake mixes or coffee syrups to increase the range of flavours. Serve them chilled or freeze them into ice lolly moulds or ice cube trays to give you more options.

Sometimes you may be asked to try individual protein supplements or a fat-free nutritional supplements (**Actagain Juce®, Altrajuce®, Ensure Plus Juice®**, **Fortijuce®**, **Fresubin Jucy®** etc.,), you should sip these slowly to give your gut more time to digest them without PERT. If you have diabetes monitor your blood glucose levels closely when taking these.

If you feel bloated with these, don't worry - this is a normal effect of taking these without PERT, but if it is affecting your quality of life, please let your dietitian know.

Phase 3: insufficient supplies and you have abdominal (tummy) symptoms

If this is not enough, and you develop symptoms such as diarrhoea, severe bloating, or urgency to need to have your bowels open, here are some other steps to try:

- If you are struggling with diarrhoea, consider taking some **loperamide / Imodium®** before your main meal. This should help to slow down your gut and reduce diarrhoea. The longer food is within your gut, the more chance more of it will be absorbed by your body. *(Please note – this is not suitable for people with CF – please contact your specialist CF Team)*
- Reduce the amount of fat in your meal to ½ of your normal the portion size of higher fat foods (Table 3). This is likely to improve some of your gut symptoms but will not mean you absorb more nutrition or prevent malnutrition, so keep a close eye on your weight and strength.
- If you eat a lot of high fibre foods – consider reducing these as very high fibre foods can bind to enzymes and make them less effective. (Table 4) Healthy eating guidelines recommend adults try to eat 30g of fibre per day. We recommend not exceeding 40g at this time.
- If you do not have diabetes, use sugary foods and drinks to increase your energy intake. Table sugar does not require enzymes to be absorbed in your gut, so Lucozade, adding sugar / honey / syrup to foods and nibbling on sugary sweets / marshmallows (not chocolate) can help keep your energy levels up. ***But*** *these do not provide any other nutrition so make sure you are having protein, vitamins and minerals from other sources.*

Phase 4: no supplies

If any one or more of these circumstances occur:

- You are unable to get hold of any PERT at all.
- You do not have enough PERT are losing weight (more than 2kg a month, or you are already underweight and losing more than 1kg a month).
- You do not have enough PERT have uncontrollable bowel symptoms that are restricting your social / work / education activities.

If you are under the care of a hospital team, contact them and see if they have sufficient supplies to issue a prescription for you – you will have to travel to the hospital to collect these if they have some available.

Inform your pharmacist that you have completely run out, so your supply can be prioritised if possible.

Try not to worry, supplies are regularly coming into the country, so this will be a short-term issue.

Contact your GP and ask for a peptide nutritional supplement to be prescribed (**Vital 1.5kcal**® or **Peptisip Energy HP**®) – you can show them this leaflet. If you are known to a dietitian – they can be contacted too, but due to the massive increase in workload this PERT shortage is generating, you are likely to get these more quickly if you go directly to your GP. These can be used instead of meals until you have your enzymes again. If you do not have diabetes, you can continue to have sugary foods and drinks alongside these. Table 2 shows how many supplement drinks you should take if you do not have any PERT at all

Table 2: Recommended doses for peptide based nutritional supplement drinks if you are unable to absorb your food.

Body weight	Supplements needed per day. (Vital 1.5kcal® or Peptisip Energy HP®)
Below 40kg (6st 4lb)	Contact a dietitian
40 – 50kg (6st 4lb- 7st 12lb)	4 x 200ml bottles = 1200kcal
50 – 60kg (7st 12lb – 9st 6lb)	5 x 200ml bottles = 1500kcal
60 - 70kg (9st 6lb – 11st)	6 x 200ml bottles = 1800kcal
70 - 80kg (11st – 12st 8lb)	7 x 200ml bottles = 2100kcal
80 - 90kg (12st 8lb – 14st 2lb)	8 x 200ml bottles = 2400kcal
Over 90kg (14st 2lb)	Contact a dietitian

This may under-estimate your needs, if you lose weight or are very active, add in one more bottle per day. If you gain weight and were not intending too – reduce by 1 bottle per day.

These do not come in a wide range of flavours, but you can add milkshake mixes or coffee syrups to increase the range of flavours. Serve them chilled or freeze them into ice lolly moulds or ice cube trays to give you more variety.

Table 3: High fat foods and their lower fat alternatives

	Reduce your portion sizes of these	Have these instead
Fats and oils	Butter, lard, Ghee, Margarine, cooking oils	Small portions of low-fat spreads Use spray on cooking oils if needed
Dairy products	Full fat milk / yoghurt Cream Crème Fraiche Cheese	Semi-skimmed or skimmed milk. Low fat yoghurts Use small amounts of grated cheese instead of slices of cheese – choose stronger cheeses to maximise taste. To increase your protein intake make skimmed milk powder up using skimmed milk and use in place of milk throughout the day
Meat and Fish	Fried foods or foods cooked in batter Skins / visible fat on meat Tinned fish, tinned in oil	Meat and fish cooked without added oil Tinned fish, tinned in spring water / brine
Plant based protein sources	Nut butters	Pulses (e.g. lentils, chickpeas, beans (note portion sizes in table 4) Quorn / Tofu – up to 100g
Fruit & vegetables	No restrictions for low fat, see Table 4 for fibre suggestions	
Carbohydrate based foods	Croissants, pastries Chips / Fried Roast potatoes	Bread, Breakfast cereals Potatoes, rice, pasta, cooked without added fat
Sauces / Condiments	Cheese based sauces Creamy sauces (bearnaise, hollandaise etc.,) Large portions of mayonnaise	Tomato based sauces, gravy, mustard, tomato ketchup, soy sauce, mint jelly, vinegar or low-fat salad dressings

Table 4: Fibre content of high fibre foods. Aim for less than 40g fibre per day

Very high fibre foods		High fibre foods			
Food	**Portion providing 10g fibre**	**Food**	**Portion providing 5g fibre**	**Food**	**Portion providing 5g fibre**
All bran®	40g	Whole wheat pitta	1 large	Weetabix®	2 biscuits
Brown pasta	250g (cooked)	Rye based crackers (i.e. Ryvita®)	4 biscuits	Shredded wheat®	2 biscuits
Baked Beans	300g	Branflakes®/ Sultana Bran®, Fruit n/Fibre®	30g bowl	Porridge / Readybrek®	Large bowl (60g oats)
Dried apricots / prunes	120g	Jacket potato with skin	1 medium	Pasta (white)	250g (cooked)
Nuts and seeds	150g	Wholemeal spaghetti	150g (cooked)	Wholemeal bread	100g
Dried lentils / chick peas /Mung beans	100g (weight before cooking)	Baked beans	150g	Quorn®	75g
Dried soya beans / red kidney beans	70g (weight before cooking)	Green beans / peas (fresh or frozen)	120g	Spinach	5 tablespoons
Desiccated coconut	70g	Sweetcorn	7 tablespoons	Avocado pear	1 whole fruit

For patients who already have a gastric feeding tube

If you have a PEG, RIG or NG feeding tube, you could take your PERT from a powdered source (Pancrex® V powder) through this tube. These can be dissolved in water and flushed down the tube, but this must be done at the time you eat. This does not work with a jejunostomy or naso-jejunal tube as the enzymes will not mix with your food. **If you have a feeding tube, discuss this option with your dietitian.**

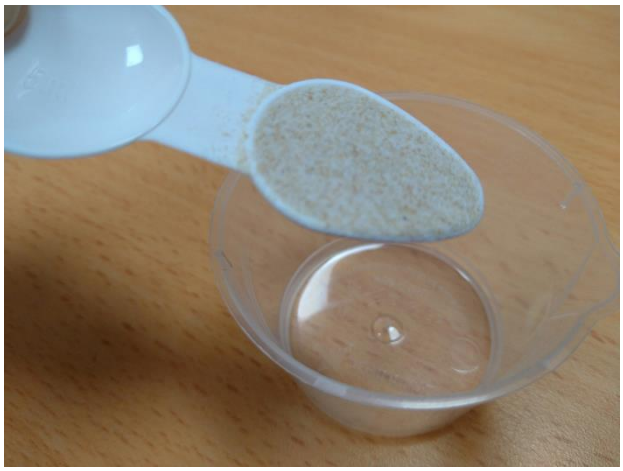
If you have CF and have any further concerns after following the relevant guidance above, please contact your CF Specialist Team/Dietitian.

Appendix 2: Guide to making up powdered enzymes

Appendix 2: Guide to making up powdered enzymes (Pancrex® V powder) for administration through a gastric feeding tube (NG,PEG or RIG) for patients who are eating.

Step 1) You will need a medicine spoon, cooled, boiled water and a pot for mixing in.

Step 2) Wash your hands. If you have eczema or sensitive skin you may wish to wear gloves for this, as Pancrex® V powder can be irritant to sensitive skin.



Step 3) The 2.5ml end of a measuring spoon measures 2g of Pancrex® V powder (50,000 units of lipase) - measure your dose of powder and place in a small bowl or cup for mixing.

Step 4) Start eating your meal.

Step 5) Once you are halfway through your meal - add 20mls cooled boiled water to the powder and mix with the medicine spoon until the powder is dissolved, don't worry if some seems to stick to the edges.

Step 6) Draw the mixture up into an enteral syringe, if some powder is stuck to the edges, squirt the mixture back into the bowl to knock it off and draw it up again.

Step 7) Flush through your feeding tube and then flush with water as normal.

You will need to do this each time you eat, the powder will only mix with the food in your stomach. If you spend more than 30 minutes eating your meal, you should take another dose.