Case Study: Lung transplantation for cystic fibrosis – A complex nutritional setting

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Introduction

Cystic fibrosis (CF) is an autosomal recessive, genetic disease that affects a variety of organs.[1,2] CF is caused by a mutation of chromosome 7 which results in defective control of sodium and chloride channels found in the cells that line a variety of organs.[1,2] This results in the production of a thick, sticky mucous that accumulates in the lungs and pancreatic ducts, as well as other mucous-producing organs.[1,2]

Management of CF includes nutritional support, pancreatic enzyme supplementation, fluidification of mucous and regular, intensive physiotherapy.[1] During periods of exacerbation, antibiotic therapy is needed.[1]

Medical advances have resulted in increased life expectancy in this patient population due to early detection through gene analysis and timely and intensive management from diagnosis.[1] The majority of deaths are caused by respiratory failure as a result of years of chronic pulmonary infections.[1]

Cystic fibrosis is the most common indication for lung transplantation in individuals under 50 years of age, accounts for 16.8% of all lung transplants in adults, and dramatically improves the survival of patients with CF with a 10-year survival rate of 45%.[3]

Case report

A 24-year-old male with CF was admitted to the intensive care unit (ICU) following a lung transplant. He had several of the described CF-related complications, namely respiratory failure, pancreatic insufficiency, CF-related liver disease and osteoporosis. The lung transplant took place on a Friday with admission to the ICU late Friday afternoon, and a full nutritional assessment took place on day three post-transplant.

Anthropometry

The patient was admitted to a private institution two months pre-transplant with a weight of 38 kg (body mass index (BMI) 13.9 kg/m^2). A percutaneous endoscopic gastrostomy (PEG) was inserted and night enteral nutrition (EN) was started. Oral nutritional supplementation (ONS) was also initiated. During the eight-week stay at the private institution, his weight increased to 43 kg (BMI 15.7 kg/m^2). He was then transferred to a tertiary hospital following the lung transplant. He maintained his weight during his post-transplant hospital stay and was discharged with a weight of 43 kg. An ideal body weight (IBW) of 63 kg (BMI 23kg/m^2) was calculated and used in calculating requirements. This is the optimal BMI range for male individuals with CF according to the current literature.[4]

Biochemistry

On admission to the ICU, the electrolyte profile and renal function were normal; haemoglobin, mean corpuscular volume (MCV) and mean corpuscular haemoglobin (MCH) were low, indicative of a hypochromic, microcytic anaemia (which could not be further characterised).[4] On day nine post-transplant albumin was low (27 g/L), and liver function tests (LFTs) were indicative of a cholestatic picture [increased alkaline phosphate (ALP) and gamma glutamyl transpeptidase (GGT)]. At discharge, albumin had improved (34 g/L) but the cholestatic picture remained (Table I).

Clinical assessment

The patient had been extubated on day two post-transplant. On the day of assessment, day three post-transplant, he was coping well on nasal cannula and was apyrexial with stable vitals. He had a soft, non-tender abdomen and had passed stools. His blood glucose levels were in a range of 7–12.5 mmol/l with a continuous infusion of insulin at 2 ml/hr (1 unit of rapid acting insulin/ml). The patient required re-intubation on day four, due to desaturation. He was extubated again on day six post-surgery and transitioned from face mask oxygen to nasal cannula. On the days following transplant, his blood glucose levels became poorly controlled with increasing doses of insulin required. At discharge on day 16 post-surgery he was

Table I. Relevant biochemistry

	Normal Values	Admission	Day 1	Day 2	Day 4	Day 6	Day 9	Day 16
Sodium	135–147 mmol/l^{-1}	141	145	142	136	140	131	142
Potassium	3.3–5.3 mmol/l^{-1}	5.5	4.4	5.0	4.4	5.9	6.6	5.6
Urea	2.6–7.0 mmol/l^{-1}	6.7	7.0	5.6	6.4	6.1	5.9	5.5
Creatinine	60–120 µmol.L^{-1}	60	65	53	78	69	71	81
Haemoglobin	14.3–18.3 mmol.L^{-1}	9.2	10.7	9.6	9.0	7.2	8.2	8.7
MCV	83.1–101.6 fL	80.2	79.1	77.5	76.4	80.8	80.2	82.5
MCH	27.8–34.8 pg	27.1	26.7	27.0	26.5	26.6	27.1	26.7
WBC	4–10 x 10^{-}	10.28	14.78	22.39	24.65	2.71	10.14	10.68
Calcium (corrected)	2.05–2.56 mmol/l^{-1}	2.17		2.04	2.06	2.04	2.04	
Magnesium	0.65–1.1 mmol/l^{-1}	0.99		0.95	1.35	0.97	0.62	
Phosphate	0.8–1.4 mmol/l^{-1}			2.22	1.38			
Albumin	35–52 g/L						27	34
DBil	0–3 umol/L						3	2
ALT	5–40 U/L						58	27
AST	5–40 U/L						41	19
ALP	40–120 U/L						275	294
GGT	0–60 U/L						234	259

WBC: White Blood count; MCV: mean corpuscular value, MCH: mean corpuscular haemoglobin; DBil: conjugated bilirubin; ALT: alanine transaminase; AST: aspartate transaminase; ALP: alkaline phosphate; GGT: gamma glutamyl transpeptidase

coping well on room air and walking without assistance. His blood glucose control improved and he was discharged without insulin or oral hyperglycaemic therapy.

Dietary management

The patient's caloric requirements were calculated using the European Society of Clinical Nutrition and Metabolism (ESPEN) guidelines of 120–150% and the United States (US) recommendations of 110–200% of energy needs for age and gender.[4] This provided a caloric range of 1 759– 3 196 kcal. Protein requirements were calculated based on ESPEN guidelines of 20% of total energy (TE) which equated to 88–160 g/day.[4]

Pre-operatively

As previously indicated a PEG was inserted for nocturnal feeds prior to the lung transplant. During this period he received the hospital full ward diet (FWD) of which he was eating approximately 50% (900 kCal, 40 g protein), 2 to 3 ONS (800 – 1 200 kcal, 40–60 g protein) and 1L semi-elemental feed via PEG overnight (1 330 kcal, 67 g protein). This provided in total 3 030 – 3 430 kcal TE and 147–167 g protein, within the calculated range. The patient was on pancreatic enzyme replacement (PERT) of 25 000 IU at meals and 10 000 IU at snacks and on supplementation of vitamin A (50 000 IU 3 times/week), vitamin D (50 000 IU once weekly), vitamin E (500 IU daily) and vitamin K (10 mg weekly), calcium (1 000 mg daily) and zinc (11 mg/day).

Post-operatively

The patient had been extubated on day one post-transplant but by day three had only taken in oral fluids and PEG feeds had not been restarted. On day three post-transplant night EN was recommenced via the PEG. A semi-elemental feed was prescribed that provided 1 330 kcal and 67 g of protein. The patient was also started on the FWD and 2 to 3 ONS (800–1 200 kcal, 40–60g protein). This provided in total 2 130–2 530 kCal and 107–127g protein, within the calculated range.

The patient however required reintubation on the afternoon of day three post-transplant and therefore was unable to take in orally. On day four post-transplant continuous EN of a semi-elemental feed was prescribed, which provided 2 660 kcal and 134 g of protein (42 kcal/kg IBW TE and 2.1 g/kg IBW protein) as per calculated requirements. The patient was extubated successfully on day six post-transplant and continued to be fed continuously until day 10 post-transplant when he began to take in orally. The feeding regimen was then changed to continuous night feeds over a 12-hour period of a semi-elemental feed which provided 1 330 kcal and 67 g protein, FWD and 2 to 3 ONS (800–1 200 kcal, 40–60g protein). He was taking approximately 75% of the FWD and managing his ONS well. Oral intake provided approximately 3 330–3 730 kcal TE and 152–172 g of protein. PEG feeds were therefore stopped. The patient maintained his weight on the FWD and ONS until discharge. The patient continued to receive PERT and vitamin A, D, E and K, calcium and zinc supplementation (dosages mentioned above) post-transplant. This regimen was also continued on discharge.

Literature review

Nutritional status, assessment and monitoring of CF patients

Undernutrition is common in CF due to a number of factors such as the underlying genetic mutation, increased caloric requirements, increased energy losses, increased essential fatty acid turnover,

a decreased dietary intake and decreased absorption of certain nutrients.[4] In this regard, an association has been reported between poor nutritional status and declining lung function in these patients, which leads to increased mortality.[4] Alternatively, there is a link between good nutritional status and improved lung function.[4]

Individuals with CF should be monitored and nutritional status assessed on a regular basis (Table II).[4] This facilitates the early identification of nutritional deficits so that treatment/interventions can be implemented.[4] In adults, weight, height and calculation of BMI should be undertaken at each clinic visit.[4,6] A multidisciplinary team should assess caloric intake, malabsorption and other factors that may influence poor weight gain.[4,6] There are a number of tools that can be used such as ONS, medications, behavioural therapy, nutritional therapies and enteral tube feeding.[4,6]

In adults (> 18 years) with CF, a focus on BMI targets is emphasised and BMI values are now recommended as the most accurate predictor of nutritional risk.[4] These targets are above 22 kg/m² in females and above 23 kg/m² in males.[4,6] It is important to note that BMI does not fully define nutritional status in these patients.[4] Increased weight from high fat mass and low lean body mass is associated with poor prognosis in CF patients.[4,6] Therefore future CF guidelines need to encompass the measurement of lean body mass in this patient population.[4,5,6] Table II summarises the nutritional status assessment methods and timing for this patient population.

Decision for intensified nutritional support

It is recommended that nutritional intervention be based on a full review of nutritional status.[4] This involves a review of adequacy and dosage of PERT and underlying medical conditions that could contribute to malnutrition.[4] BMI cut-offs are used to determine whether progressive and more specialised nutrition intervention is necessary (Table III).[4]

EN should be considered in individuals with a persistently low BMI (< 18.5) and/or ongoing weight loss (> 5%), where diet modification and/or ONS have failed to improve nutritional status.[4] EN can be administered as continuous infusions or as bolus feeds.[4] They can be given over a 24-hour period or as nocturnal feeds.[4] Nocturnal feeds can be beneficial as these make it easier for patients to eat a high calorie diet during the day.[4]

There is not enough evidence in the literature to recommend a specific type of enteral formula (polymeric, semi-elemental, elemental).[4,6] According to ESPEN guidelines, most individuals tolerate high calorie (1.5–2 kcal/ml) polymeric formulas well. In individuals who do not, an elemental or semi-elemental formula can be used.[4]

There is insufficient evidence regarding the administration of PERT with EN.[4,6] The most commonly used method is to give PERT at the beginning and end of the feed infusion and in the middle of the night if necessary.[4] Bolus feeds may require more frequent and higher doses of PERT.[4,6]

Parenteral nutrition is rarely used in CF patients and should only be used where enteral feeding is contraindicated.[4,6]

CF-related complications with nutritional consequences

Pancreatic insufficiency

Pancreatic insufficiency (PI) occurs in a large number of CF individuals.[4] PI is defined as a decrease in digestive enzymes and is associated with malabsorption.[4] PERT is recommended for all patients with PI.[4,5] PERT adequacy should be determined clinically by monitoring nutritional status, symptoms of malabsorption, excessive appetite and poor weight gain or weight loss.[4,5] These parameters should be monitored every six months in adults with CF.[4] Table IV shows dosage guidelines for initiation and continuation of PERT.

Table II. Assessment of nutritional status for adult patients with cystic fibrosis[4]

	Assessment method and timing
Weight monitoring	• Measure weight at each clinic visit. • Clinic visits at least every three months.
Nutrition monitoring	• Annual nutritional review including blood tests (full blood count, iron status, plasma fat-soluble vitamin levels, plasma or serum phospholipid fatty acid patterns, serum liver function and electrolyte measurements). • Monitor pancreatic enzyme replacement therapy (PERT) and vitamin levels three to six months after initiation or change in dosage. • Consider the following markers of risk for malnutrition: ◦ Measurement of pulmonary function ◦ Annual assessment of pancreatic function (by faecal pancreatic elastase-1 determination) in pancreatic-sufficient individuals. More frequent tests if nutritional status is poor/not improving. ◦ Assessing for PERT need or adequacy of treatment every six months by monitoring nutritional status. Use a 72-hour faecal fat measurement and the calculation of the coefficient of fat absorption in patients whose nutritional status is questionable.
Dietary review	• A dietary review (which includes questions regarding adherence to advice) should be conducted at least every six months.

Table III. Consensus guidelines on nutrition intervention in cystic fibrosis[4]

Nutritional status	Intervention	Decision point for intensified nutrition support Adults (> 18 years)
Normal nutritional status	Preventative nutritional counselling	BMI: 18.5–22 (for females); 18.5–23 (for males), or no weight loss
Impaired nutritional status	Diet modification and/or oral nutritional supplements	BMI < 18.5, or weight loss of 5% in previous two months
Persistent undernutrition	Enteral tube feeding	Persistently low BMI (< 18.5), or ongoing weight loss of > 5%

*BMI = Body Mass Index

Table IV. Pancreatic enzyme replacement therapy (PERT) dosage guidelines[3]

Age	Suggested dosage
Children > 4 years and adults	Start at 500 U lipase/kg/meal and titrate upwards to a maximum dose of: -1 000–2 500 U lipase/kg/meal, or -10 000 U lipase/kg/day, or -2 000–4 000 U lipase/gram dietary fat take from all fat-containing meals, snacks, drinks

U: Units

Bone disease

Osteoporosis and osteopenia are common in many adolescents and adults with CF.[4,5] Low bone mineral density is associated with decreased ability to work, increased risk of fractures, severe lung disease, essential fatty acid (EFA) insufficiency and decreased fat mass.[4] Poor nutritional status and deficiencies of vitamin D, K and calcium are all indicators of nutritional risk.[4] CF individuals with osteopenia or osteoporosis should be educated on increasing their intake of dietary sources of calcium and EFAs. Supplemental calcium, vitamin D and vitamin K may also be warranted.[4,5] The use of bisphosphates can be considered on an individual basis.[4] Factors such as bone mineral density, history of fractures and transplant status should be considered.[4] It is also recommended that individuals with CF exercise regularly and include weight-bearing exercise as this has been shown to improve bone mineral density.[4]

CF-related diabetes

Many individuals with CF develop CF-related diabetes (CFRD) due to decline in the endocrine function of the pancreas over time.[4] CFRD is becoming more prevalent as individuals with CF are living longer and the prevalence of CFRD increases with age.[4] CFRD negatively affects lung function as there is an increase in bacterial colonisation of the lungs with increased blood glucose levels.[8] It is recommended that CF individuals over the age of 10 years are screened for glucose tolerance annually.[4,5] The American Diabetes Association, Cystic Fibrosis Foundation and the Paediatric Endocrine Society advise a high calorie diet where carbohydrate intake is individualised and regularly monitored to obtain good glycaemic control.[4,5] A higher protein intake and high fat intake that includes EFAs is recommended.[4,6] The use of artificial sweeteners should be limited as these can prevent adequate caloric intake.[8] Education should also include advice on insulin therapy and aerobic exercise.[8] Insulin is recommended as the treatment of choice as there is insufficient evidence for the use of oral hypoglycaemic agents in CFRD.[4,5]

Liver disease

A large number of individuals with CF will develop liver disease ranging from mild biliary fibrosis to end-stage cirrhosis.[5] Cystic fibrosis related liver disease (CFLD) usually presents before 20 years of age.[5] Some individuals with CFLD develop portal hypertension, which can cause variceal bleeding and hepatic failure.[4,5] There is an association between liver disease and hepatic steatosis in CF individuals.[4] ESPEN therefore recommends supplementation with fat-soluble vitamins and EFAs. Older individuals with CF may develop liver failure.[4] There is an association between impaired liver function and declining lung function.[4] All individuals with CF should receive routine physical examinations and liver enzyme tests should be undertaken on an annual basis.[5]

Gastrointestinal complications

There are a number of gastrointestinal complications associated with CF.[5]

Gastroesophageal reflux disease (GORD) affects approximately 30% of individuals with CF.[5] Care providers should be aware of signs and symptoms of GORD and appropriate diagnostic testing and treatment should be provided.[4,5]

Constipation is frequently experienced by CF individuals.[5] Treatment usually involves education on adequate diet and hydration and the use of stool softeners or laxatives.[5] Enemas are not usually needed.[5]

Fibrosing colonopathy (FC) is a rare complication in CF individuals and is usually caused by overuse of PERT.[5] It is characterised by thickening of the bowel wall and generalised narrowing of the bowel lumen.[5] The ascending colon is most commonly affected but the entire colon can be affected.[5] Using the appropriate dosage of PERT, not increasing dosages unnecessarily and not exceeding a total enzyme dosage of 10000 lipase units/kg/day to prevent FC are the only recommendations to prevent FC.[5]

CF individuals with small bowel bacterial overgrowth (SBBO) experience symptoms such as abdominal pain, nausea, excessive flatulence and malabsorption (with adequate PERT).[5] Diagnosis should be made based on a clinical trial of metronidazole to treat the bacterial overgrowth.[5] A referral to a gastroenterologist is necessary.[5]

Energy and protein requirements

CF patients have higher energy needs than the general population.[4] This is due to various factors such as chronic inflammation, impaired lung function, malabsorption and the presence of acute respiratory exacerbations.[4] ESPEN guidelines recommend that adult CF patients should receive 120–150% of energy requirements for same-age healthy populations to maintain BMI targets.[4] In comparison, US guidelines recommend that CF adult patients should receive 110–200% of energy requirements for same-age healthy populations to maintain BMI targets.[4]

Protein needs are estimated to be higher in individuals with CF, compared to the general population. Current recommendations are that protein should be 20% or more of TE intake.[4] Further studies are needed to determine more definitive guidelines for protein.[4]

Essential fatty acids (EFAs)

Humans are unable to synthesise EFAs and therefore they have to be obtained via dietary sources.[4,6] EFA deficiency is more common in CF individuals with pancreatic insufficiency due to the malabsorption of fat.[4,6] EFA deficiency may present as dermatitis, alopecia and/or thrombocytopaenia.[6] Low DHA (docosahexaenoic acid) and high AA (arachidonic acid) is associated with decreased bone mineral

density among children and young adults with CF.[4] Consideration of EFA deficiency and treatment is recommended by the Cystic Fibrosis Foundation in individuals with poor weight gain or weight loss.[6] A few studies have demonstrated that increased intake of EFAs may improve lung function.[4,6] Some studies have shown that supplementation with omega-3 fatty acids may deliver anti-inflammatory benefits with few negative side-effects.[4,6] At present, there is insufficient evidence to make specific recommendations regarding dietary supplementation of EFAs.[4,6]

Electrolytes, minerals and trace elements

Individuals with CF have increased requirements for certain electrolytes, minerals and trace elements compared with non-CF individuals.[4,6]

Excessive salt loss through sweat can cause decreased levels of sodium.[4,6] Most adults with CF will obtain adequate sodium from a Western diet that includes processed foods.[4] However certain conditions such as fever, exercise and hot weather can result in increased sweat production and depleted levels.[4,7] Supplementation is recommended in these situations and can be given in the form of additional salty foods, sodium chloride capsules or vials.[4,7] Individuals in temperate climates or while exercising may require extra sodium supplementation in the form of salted sports drinks.[4,7]

Patients with CF often have low levels of calcium.[4,7] This is due to a combination of decreased intake of dietary sources of calcium and a deficiency of vitamin D.[4] Gastrointestinal malabsorption, due to PI, that is not fully corrected by PERT may also cause decreased calcium levels.[4] Calcium intake should be assessed annually or more often if necessary.[4,7] Individuals with suboptimal levels of calcium should be counselled on increasing their intake of dietary sources of calcium.[4] If dietary intake remains low, a calcium supplement is recommended.[4,7]

Iron deficiency is common in patients with CF with an estimate that more than 50% of stable adult CF patients have an iron deficiency.[4,7] There are a number of factors that can contribute to iron deficiency, such as chronic infection and inflammation, inadequate dietary intake, malabsorption and blood loss. It is recommended that patients are only started on iron supplementation if a deficiency persists once underlying inflammation and infection have been treated and resolved.[4,7] Individuals with CF should be routinely monitored for anaemia by first obtaining serum iron levels.[4] If serum iron levels are low it is important to distinguish between iron deficiency anaemia (IDA) and anaemia of chronic disease/inflammation (ACD/AI).[4] In cases of ACD/AI, supplementation is cautioned as iron overload may exacerbate the oxidative stress from chronic inflammation in CF.[7] Table V shows how to differentiate between IDA and ACD/AI.[4,7]

There are a number of studies that have reported altered zinc status in CF individuals.[4] Zinc plays an important role in both immunity and growth.[6] Plasma zinc levels are not a sensitive indicator of zinc status as it is influenced by a variety of conditions.[4] Zinc supplementation is recommended in CF individuals at risk of a zinc deficiency.[4] Factors such as increased number of infections due to impaired immune function, eye and skin lesions, hair loss, diarrhoea, taste abnormalities, delayed wound healing and anorexia may be indicative of a zinc insufficiency.[4]

Fat-soluble vitamins

Malabsorption due to PI can result in deficiencies in fat-soluble vitamins (Table VI).[4,7] Some CF individuals who are pancreatic sufficient have also been shown to be at risk for fat-soluble vitamin deficiencies.[4] It is recommended that plasma levels of fat-soluble vitamins are measured annually or more often if necessary, in CF individuals without proven PI.[4] For PI individuals it is recommended that plasma levels of fat-soluble vitamins be measured at initiation of PERT and vitamin supplementation and three to six months later or if there were changes in the supplementation regimen.[4] Fat-soluble vitamin supplements should be taken with a high fat food and PERT.[4,7]

Vitamin A deficiency has been reported in 10–40% of CF individuals.[4,7] Suboptimal vitamin A levels are associated with declining lung function, increased exacerbations and worsened clinical status.[4,7] Vitamin A toxicity is an important factor to consider when deciding on supplementation.[7] Vitamin A supplementation should aim to achieve the normal range of serum retinol concentrations for healthy individuals.[4] Beta-carotene is a precursor to vitamin A and is controlled by a negative feedback mechanism and is therefore safer to use than retinol (Table VI).[4]

Vitamin D has an important role in intestinal calcium absorption and a deficiency can result in decreased bone mineral density (BMD).[4,7] A major source of vitamin D is sunlight, however exposure is often limited.[4,7] Serum 25-hydroxy vitamin D (25(OH)D) is used to monitor vitamin D status.[4,7] Supplementation is recommended to maintain serum 25(OH)D above 20 ng/mL (50 nmol/L).[4] Vitamin D3 (cholecalciferol) is recommended over vitamin D2.[4] It is recommended that serum 25(OH)D is monitored annually, preferably at the end of winter.[4]

Alpha-tocopherol is the major compound of vitamin E.[4] Vitamin E deficiency can cause a variety of serious side-effects such

Table V. How to differentiate between different forms of anaemia[4]

	Iron deficiency anaemia (IDA)	Anaemia of chronic inflammation	Both forms of anaemia
Serum iron	Below normal	Below normal	Below normal
Serum ferritin	Below normal	Above normal	Varies
Total iron binding capacity	Above normal	Below or normal	Varies
Transferrin saturation (%)	Below normal	Below normal	Below normal

as neuromuscular degeneration, haemolytic anaemia and both retinal and cognitive deficits.[4,7] CF individuals often have increased requirements during periods of pulmonary exacerbation; this increases oxidative stress.[4] Studies that demonstrated that vitamin E supplementation increases serum vitamin E levels failed to show a direct clinical benefit.[4,7]

A number of studies have shown that CF individuals often have suboptimal levels of vitamin K and those with CFLD are almost always deficient.[4,7] Vitamin K plays an essential role in blood clotting and also contributes to bone health.[4,7] The measurement of vitamin K is problematic in practice and tests are expensive and therefore not routinely used.[4,7] The measurement of prothrombin time is insensitive and only elevated in a severe deficiency.[4,7] Regular supplementation of dietary vitamin K_1 is recommended.[4] Currently there is insufficient data available to determine the appropriate dosage of vitamin K.[4]

Table VI. Recommended dosages for fat-soluble vitamin replacement in cystic fibrosis[4]

	Starting dose	Maintenance/ maximum dose
Vitamin A	Beta-carotene 1 mg/kg/day for 12 weeks (Maximum 50 mg/day).	10 mg/day
Vitamin D	800 IU/day	2 000 IU
Vitamin E	100–400 IU	
Vitamin K	1–10 mg/day	

Probiotics

The use of probiotics in improving health outcomes related to respiratory, gastrointestinal and nutrition in CF is a new topic that is being investigated.[2]

Two randomised controlled trials (RCT) showed that probiotics resulted in a reduction in pulmonary exacerbations (PE).[2] Two pilot studies investigated the role of probiotics in reducing the duration and severity of PE but showed no clinical benefit.[2] Two RCT investigated the effect of probiotics on the number and duration of hospital admissions for PE.[2] The results were inconclusive for the effect of probiotics on hospital admissions and showed no effect on duration of hospital stay.[2]

In terms of improvements in gastrointestinal and nutritional outcomes the results are either currently inconclusive or showed no significant functional improvement.[2]

Conclusion

This patient presented with a combination of CF-related complications that were managed or improved through nutritional intervention. Following the current guidelines and recommendations as well as regular monitoring of nutrition-related complications dramatically improves the quality of life and prognosis of CF patients.

The patient was discharged on post-transplant day 16 and received ONS for home use (500 kCal TE, 14 g protein per day). He is being followed up as an outpatient and has gained 4.5kg since discharge. He has also started exercising under physician supervision and is able to cycle 2 km. His nutritional regimen will be monitored and adjusted as his activity level increases to ensure adequate weight gain and maintenance.

References

1. Samano MN, Pêgo-Fernandes AK, Fonseca Riberio K, et al. Lung transplantation in patients with cystic fibrosis. Transplanation Proceedings. 2013;45:1137-1141.
2. Ansderson JL, et al. Effect of probiotics on respiratory, gastrointestinal and nutritional outcomes in patients with cystic fibrosis: A systematic review. Journal of Cystic Fibrosis. 2017;16:186-197.
3. Snell G, et al. The evolution of lung transplantation for cystic fibrosis: A 2017 update. Journal of Cystic Fibrosis. 2017;16:553-564.
4. Turck D, et al. ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children and adults with cystic fibrosis. Clinical Nutrition. 2016:1-21.
5. Smyth AR, et al. European Cystic Fibrosis Society Standards of Care: Best Practice Guidelines. Journal of Cystic Fibrosis. 2014;13:S23-S42.
6. Sullivan JS, Mascarenhas MR. Nutrition: Prevention and management of nutritional failure in cystic fibrosis. Journal of Cystic Fibrosis. 2017;16:S87-S93.
7. Li L, Somerset S. Dietary intake and nutritional status of micronutrients in adults with cystic fibrosis in relation to current recommendations. 2016;35:775-782.
8. Waugh N, Royle P, Cragie I, , et al. Screening for cystic fibrosis related diabetes: a systematic review. Health Technol Assess. 2012;1-179.