School of Public Health

DEVELOPMENT, IMPLEMENTATION AND EVALUATION

OF A NUTRITION EDUCATION

AND BEHAVIOUR PROGRAM

FOR CHILDREN WITH CYSTIC FIBROSIS

by

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This thesis is dedicated to the memory of my father,

Kenneth Richard Hastie, 1931 to 1991,

who, together with my mother,

lovingly and continuously supported

the ventures I pursued in life which ultimately led to

the program and research described in this manuscript.

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Publications related to this work

(Appendix 8)

Stapleton D, Tunnecliffe L, McGuiness D, Sherriff J and Sly P. Development of a nutrition and behaviour intervention program: Go and Grow with CF. Aust J Nutr Diet 1998;55(3):130-137.

Stapleton D, Gurrin L, Zubrick S, Silburn S, Sherriff, J and Sly P. What do children with CF and their parents know about nutrition and enzymes? J Am Diet Assoc 2000;100(12):1494-1500.

Stapleton DR, Gurrin LC, Zubrick SR, Silburn SR, Sherriff JL and Sly PD. The effect of 'Go and Grow with CF' on nutrition and pancreatic enzyme knowledge of children with cystic fibrosis. Aust J Nutr Diet 2001;58(3):164-168.

Literature review

The literature that was extensively reviewed for this thesis
included publications up to 1998

ABSTRACT

Background: Cystic fibrosis (CF) is a genetically inherited disease which adversely affects the respiratory and gastrointestinal systems. Malnutrition is a major clinical problem in individuals with the disease. Nutritional interventions are warranted as improvements in nutritional status could improve the rates of morbidity and mortality associated with the disease. The review of the literature indicated the need to develop a behavioural-based nutrition prevention program in order for children to achieve CF dietary requirements and appropriate pancreatic enzyme replacement therapy.

Methods: The intervention program, *Go and Grow with CF*, and nutrition and pancreatic enzyme knowledge and self-management questionnaires were developed for children with CF and their carers as part of this thesis. Social learning theory constructs which particularly assist children in achieving desirable behaviours were applied during the development of the *Go and Grow with CF* program. The program consisted of workshops and a home-based course.

Fifty eight children with cystic fibrosis, aged 2 to 11 years, and their carers participated in a clinical trial that was designed to assess the effects of the *Go and Grow with CF* pilot program on knowledge, self-management, behaviour, dietary intake and body composition, using anthropometry. Process evaluation was conducted on the pilot program and on the clinic-wide implementation of the revised *Go and Grow with CF* program. The revised program included the Australian Pancreatic Enzyme Replacement Therapy Guidelines and the effects of fat-based dosing were assessed with a cohort of 29 children with CF-related pancreatic insufficiency aged 1 to 13 years.

Results: Similar to the process evaluation of the pilot program, 100% of carers who completed the revised home-based course indicated that they would recommend *Go and Grow with CF* to other families with a child who has CF. The objective assessment of knowledge indicated a significant improvement in children's knowledge in the short-term. There were no statistically significant improvements in

any of the other parameters assessed. The lack of significant improvements in self-management, behaviour, dietary intake and anthropometry may have been because the program had no effect, the parameters assessed or the instruments used (particularly the questionnaires) were not sufficiently sensitive, the sample size (which was determined by the CF population available) was too small or the duration of the intervention and follow-up was too short.

Conclusion: Carers' unanimous recommendation of *Go and Grow with CF*, together with high levels of perceived learning, reported increase in confidence and improvement in children's knowledge in the short-term, indicate the benefits of the program.

Although there was no statistically significant improvement in the anthropometric measurements after the intervention, the extensive data obtained during this study suggest that measurements of height and weight may underestimate the presence of poor nutritional status. It is likely that comprehensive assessments of body composition of children with CF would be useful in detecting mild degrees of malnutrition and in providing information about the effects of nutritional status on morbidity and mortality associated with the disease.

Fat-based pancreatic enzyme replacement therapy dosing warrants further investigation given that parents had a strong preference for this method and that fat absorption remains abnormal in the majority of individuals who have pancreatic insufficiency. Evaluation of all pancreatic enzyme replacement therapy dosing methods are needed and this research suggests that dose should be assessed on a meal and snack basis, rather than just on daily intake, in order for levels of adherence to be examined.

The apparent absence of a long-term effect of a single exposure to the program on knowledge suggests that regular, ongoing education and counselling is required by families to reinforce aspects related to the child's current stage of development and disease status.

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ABBREVIATIONS

AM	Adipose mass
AMA	Arm muscle area
BPFAS	Behavioural Pediatrics Feeding Assessment Scale
COGRO	Coquitlam growth study
CF	Cystic Fibrosis
FFB	Faecal fat balance
FFQ	Food frequency questionnaire
$FEF_{25\% \, to \, 75\%}$	Forced expiratory flow from 25% to 75% of vital capacity
FEV_1	Forced expiratory volume in 1 second
FVC	Forced vital capacity
GPHNS	Geelong Public Health Nutrition Survey
IU	International units
MUAC	Mid-upper arm circumference
MM	Muscle mass
NCHS	National Centre for Health Statistics
PERT	Pancreatic enzyme replacement therapy
%fat	Percent body fat
%FFE	Percent faecal fat excretion
%IBW	Percent ideal body weight
RDI	Recommended daily intake
$\Sigma 6SF$	Sum of 6 skinfolds
USA	United States of America

1. CHAPTER ONE: INTRODUCTION

1.1 Significance of the study

Malnutrition continues to be a major clinical problem in children with cystic fibrosis (CF) as indicated by the retardation in weight gain and linear growth of most patient populations. For example, the 1995 United States of America (USA) CF Foundation patient registry indicated that 25% of children with CF were <5th percentile for height and 20% were <5th percentile for weight (1). It is possible that the prevalence of sub-optimal nutritional status in CF is underestimated due to the heavy reliance on measures of height and weight (2).

Malnutrition has been associated with increased morbidity and mortality in CF (3, 4). The maintenance of energy balance is thought to be challenged by a combination of factors. Energy deficits may arise as a consequence of CF-related increases in energy expenditure, increases in gastrointestinal losses and decreases in oral intake. Advances in pancreatic enzyme replacement therapy (PERT) and dietary recommendations over the past two decades have resulted in significant increases in the fat intake and absorption of individuals with CF, but numerous issues (Section 2.4) continue to limit the overall oral intake of children, such that elevated requirements are rarely met.

Enteral-tube feeding has been successful in enhancing the nutritional status of malnourished individuals with CF and behavioural therapy programs have assisted parents in dealing with meal-time problems. However, it has been difficult to prevent or at least minimise problem behaviours and malnutrition due to limited availability of resources that equip families with the skills necessary to achieve optimum dietary and PERT management of children with CF.

The intention of this study was to develop and assess the effects of an intervention program designed to facilitate children with CF and their carers in the process of translating knowledge into desirable nutrition and PERT behaviours. A carer was defined as the adult most responsible for the child's nutrition and PERT needs. It was envisaged that both the families of children with CF and the dietitians caring for these children in centres throughout the world would benefit from the study as no program like the one presented in this thesis existed.

1.2 Research questions

Two research questions were addressed.

1. Will a nutrition intervention program that is based on a model of behavioural change, and specifically designed for children, facilitate the translation of knowledge into behaviour and action, and thereby improve nutritional status?

2. Will matching PERT dose to dietary fat intake improve fat absorption in children with CF?

1.3 Objectives of the research

The primary objectives of the initial study were:

1. To develop and implement a behavioural-based nutrition program.

2. To increase the nutrition and PERT knowledge of children with CF aged 6 to 11 years and carers of 2 to 11 year olds.

3. To facilitate the adoption of nutrition and PERT self-management skills by children with CF aged 6 to 11 years and carers of 2 to 11 year olds.

4. To enhance the number and range of strategies used by carers in coping with dietary- and PERT-related behavioural problems.

5. To decrease dietary- and PERT-related behavioural problems of children with CF aged 2 to 11 years as reported by carers.

Secondary objectives of the initial study were:

6. To assess changes in dietary energy and nutrient intakes using a food frequency questionnaire.

7. To determine if a range of anthropometric measurements can detect early signs of malnutrition in children with CF.

8. To assess changes in body composition using skinfold and circumference measurements.

The specific objectives of the subsequent study were:

9. To evaluate the process of implementing the revised intervention program.

10. To assess the effects of fat-based dosing on fat absorption in children with CF.

2. CHAPTER TWO: LITERATURE REVIEW

This review reports on the nutritional issues surrounding children with CF and strategies for improving oral intakes and absorption as described in the nutrition, health education, psychology and CF literature.

2.1 Incidence and survival

CF is the most common lethal genetic disease in the Caucasian population with 1 in 25 of the population being carriers and an incidence of approximately 1 in 2 500 live births. The most common presentations of the disease are failure to thrive, malabsorption and respiratory problems. Survival largely depends on the rate of progression of lung disease, with most individuals with CF dying of cardio-respiratory failure. The mean survival age varies slightly between countries and genders, being 30 to 40 years for males and 25 to 28 years for females (5, 6). The cause of the gender difference has been extensively investigated, but is still unknown. The projected life expectancy of children born with CF in the late 1990s may be much greater than in previous years, due to marked advances in treatment of the disease and early diagnosis with neonatal screening programs.

2.2 Clinical features of CF

In CF, an autosomal recessive single gene defect causes unusually thick mucus to be secreted from exocrine glands. Mutations of the cystic fibrosis transmembrane conductance regulator gene, which was identified on chromosome 7 in 1989, limit chloride secretion and possibly increase sodium absorption in airway epithelia, which subsequently alters the consistency of airway secretions. Abnormally large amounts of very viscous mucus are secreted, such that mucociliary clearance is impaired and the small airways become plugged with mucus (7). The cystic fibrosis transmembrane conductance regulator defect may also enhance bacterial adherence to

epithelial cells, which leads to persistent infection and an associated inflammatory response (8).

Clinical features of CF most often include chronic obstructive lung disease with predominant airway involvement and recurrent infections and abnormally high sweat sodium and chloride levels, due to failure of salt reabsorption in sweat gland ducts. Other features may include steatorrhea and azotorrhea due to exocrine pancreatic insufficiency, intestinal obstruction in the neonate or older patient, failure to thrive and malnutrition, chronic sinusitis, cirrhosis of the liver, clubbing of the extremities, osteopenia, episodic arthritis and infertility. Most of the signs of malnutrition in CF (weight deficits, growth retardation, delayed bone age, loss of adipose tissue, muscle wasting, hypoalbuminaemia, oedema, anaemia, bruising, bleeding, immune and respiratory dysfunction, developmental delay and delayed puberty) can be related to a protein-energy deficit or the malabsorption of essential nutrients (9).

Those with the disease have two abnormal copies of the defective gene. The most common mutation is at position 508. The correlation between genotype and phenotype is not strong and the course of the disease can be quite different in patients even with the identical genotypes. The high level of variation in disease expression, in both the severity of the symptoms and the rate of decline in pulmonary function, is thought to be because of the numerous mutations of the CF gene (in excess of 600 mutations have been identified) and varying environmental influences. Although the genetic defect of CF is increasingly understood there is no specific therapy available and treatment is limited to dealing with the complications of the disease. The complexity and progressive nature of CF means that optimal management may not always lead to good medical outcomes.

Episodes of complications and serious illnesses not only affect health status but also the patient's and family's psychosocial functioning. A family with a chronically ill child is required to integrate numerous therapies into usual daily routines, in addition to maintaining normal family life. Families not only have to cope with the disruption that CF treatments cause to relationships, but also with the terminal nature of the

disease. Management of this multi-system disease most often involves a comprehensive, multidisciplinary team approach, with the aim being to improve the duration and quality of life of individuals who have the disease. Together with respiratory treatments, nutrition is increasingly viewed as an integral part of the care of individuals with CF and programs are sought to prevent malnutrition.

2.3 Malnutrition

Malnutrition is a major clinical problem in CF, with significant retardation in weight gain and linear growth failure during childhood being recognised among most patient populations. The 1995 United States of America (USA) CF Foundation patient registry indicated that 25% of children with CF are <5th percentile for height and 20% are <5th percentile for weight (1). Morison et al (10) report that the mean height and weight of children with CF in the first decade of life in the United Kingdom are about 0.5 standard deviations below those of the general population. These data are from cross-sectional surveys of patients attending CF specialty centres and may not include the entire CF population in the area (1, 10). Australian studies report mean z-scores of -0.3 for weight and between -0.8 and -0.3 for height (11, 12). Regardless of the incompleteness of the international and national data available, the results support the notion that progressive or episodic deterioration in growth and nutritional status continue to occur in individuals with CF throughout childhood and adolescence, and that nutritional deficits are more prominent in infants, older children and in females (11-14).

With regard to infants, reviews of the literature (10, 15) have identified that there is controversy regarding whether or not birth weight is normal in those with CF. If birth weight is low then it could contribute to growth deficits in young patients and could also be indicative of an intrinsic constraint on growth in CF (10). As the disease progresses with age, inadequate weight gains are observed in older children (16). The higher mortality rate for females with CF (17) is poorly understood; it is possible that the aetiology is linked to nutritional status. The cross-sectional survey by Morison et al (10) demonstrates the differences that exist in growth parameters

between genders. Mean weight z-scores of 0 to 10 year old boys with CF ranged from -0.25 to -0.5 for each year of age and most of the mean values for each year of age of the girls approximated 0.5 standard deviations below the reference mean (10). However, it is not clear from the publication whether the difference between genders is significant. Further assessment of the gender effects of malnutrition on survival requires comprehensive assessments of nutritional status.

There is the possibility that the extent of malnutrition in individuals with CF is underestimated due to the heavy reliance on assessments of growth (height and weight). The continuing reliance on height and weight as indicators of nutritional status may be because CF was traditionally a disease of childhood and these measures are routinely obtained from children in the clinical setting. Use of some of the indices derived from measures of height and weight, such as the percent ideal body weight (%IBW) index (18), may further limit the identification of individuals at nutritional risk. The %IBW index does not detect stunting in growth as it is an assessment of weight relative to height. In contrast, the z-score is more useful as height and weight can be separately compared to a reference population. Thus, a comparison by Anthony et al (11) of the growth of children with CF and their siblings using mean %IBW index values suggested that the two groups were of similar nutritional status, but the age- and gender-adjusted standard deviation- or z-score values indicated that the children with CF were shorter and lighter than their siblings. The measures used to assess nutritional status in CF need to be extensively reviewed in order to ensure that deteriorations can be detected early, so that malnutrition is prevented or at least minimised (18).

Malnutrition was initially accepted as an inherent consequence of CF and the result of a physiological adaptation to advanced pulmonary disease. This was until the 1980s when Corey et al (3) observed a positive association between better growth, less severe pulmonary disease and higher rates of survival. CF clinic patients from Toronto were found to have a nine year survival advantage over those attending the Boston CF clinic. The improved rate of survival for the Toronto clinic was associated with better growth parameters, with patients being taller and heavier than

their Boston counterparts. The differences in height and weight were attributed to differences in the dietary regimens advocated by each clinic as all other management techniques were similar between the two CF centres. The Boston CF clinic adhered to the universal low fat diet recommendation, that was instituted in an attempt to alleviate the problem of steatorrhea in patients with pancreatic insufficiency. The Toronto CF clinic recommended an unrestricted fat diet with an accompanying increase in pancreatic enzyme doses, as the difficulty of achieving a high energy intake with minimal fat had been recognised. The positive association between the higher fat diet, better growth, improved pulmonary function and higher life expectancy, together with refinements in pancreatic enzyme preparations, resulted in marked changes to the nutritional management of CF. CF centres were advised not to restrict dietary fat (3) and many started to advocate a high fat diet in order for individuals with CF to be more likely to achieve elevated energy requirements. The subsequent observation of an association between weight gain and improvement or stabilisation of pulmonary function, in malnourished patients receiving increased energy intakes through enteral-tube feeding, has provided further support for optimising nutritional management in CF (19-21).

Although the degree of underweight closely correlates inversely with survival in CF (4), it is not known whether low weight is a cause of declining pulmonary status, or merely a marker of the progression of the disease (22). Figure 2.1 illustrates the complexity of the inter-relationship between the CF disease process, infection, lung disease and malnutrition (9). Recurrent chest infections, and the accompanying anorexia and increase in energy expenditure, can adversely affect nutritional status and growth. Conversely, malabsorption, specific nutritional deficiencies and protein-energy imbalance may result in altered pulmonary defence mechanisms, decreased exercise tolerance and altered pulmonary muscle function (23). Although the aetiology of malnutrition in CF is unclear, the link with life expectancy underlines the importance of addressing the factors that influence nutritional status.

Figure 2.1

Pathogenesis of energy imbalance in cystic fibrosis
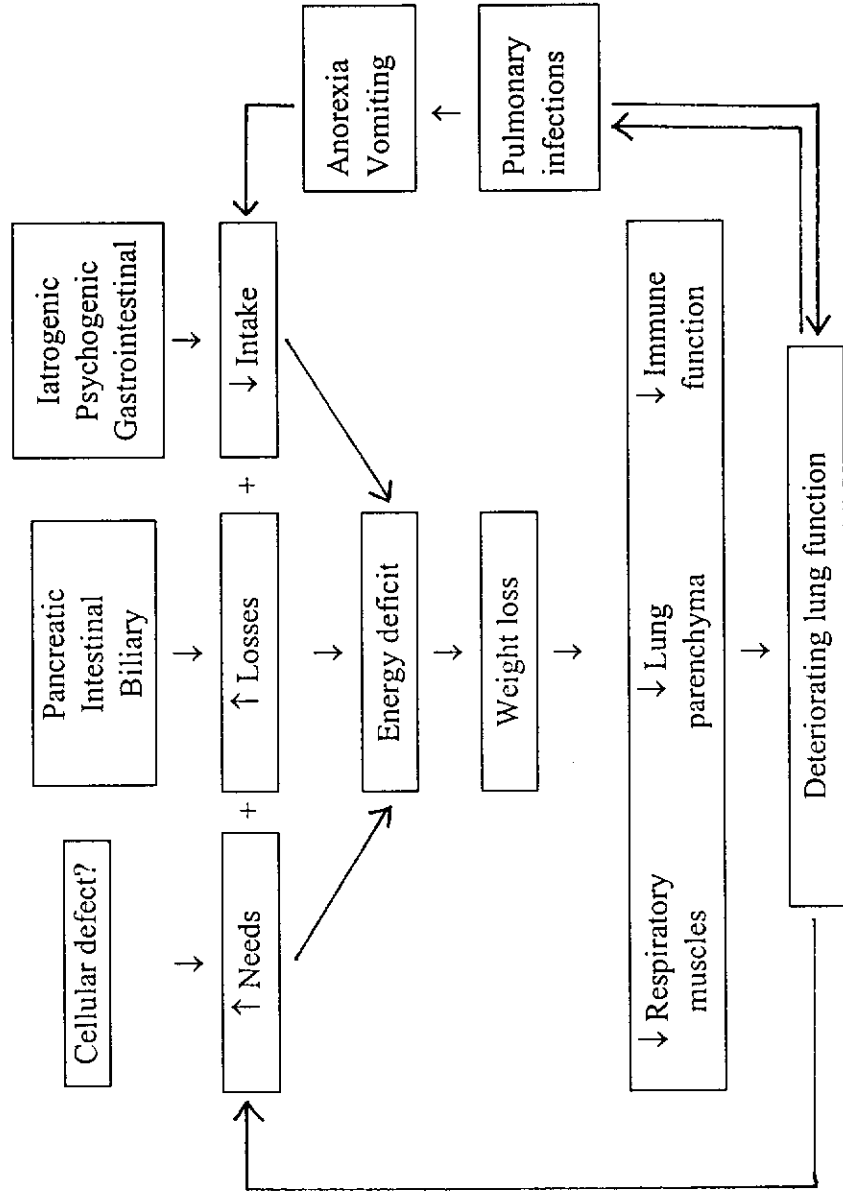
From Forstner and Durie (9)

2.4 Aetiology of malnutrition in CF

Malnutrition in individuals with CF is mostly due to a complex combination of factors that challenge the maintenance of energy balance (14, 24). An energy deficit arises when there is an imbalance between energy needs and intake and is determined in CF by three factors, namely increased expenditure, increased losses (both before and after absorption) and decreased intake (Table 2.1) (25).

Table 2.1

Energy imbalance in cystic fibrosis

Increased expenditure	Increased losses	Decreased intake

Pulmonary disease
? primary defect
	Intestinal
	    Pancreatic insufficiency
	    Bile salt metabolism
	    Hepatobiliary disease
	    Gastroesophageal reflux
	
	Sputum
	
	Urinary
	    CF-related diabetes
		Reduced intake
		    Anorexia
		    Feeding disorders
		    Depression
		    Oesophagitis
		    Intestinal obstruction
		    Food preferences
		    Cytokine release
		    Abdominal pain
		    Psychosocial aversion
		        to fat

Adapted from Wilson & Pencharz (25)

In children with CF, the maintenance of adequate growth and nutritional status is often more difficult as a positive energy balance is required to meet the energy cost of synthesising new tissue and the energy content of the new tissue deposited. Although only a crude estimate, it is often suggested that individuals with CF require between 120% and 150% of the normal recommended daily intake (RDI) of energy for age and gender to compensate for elevated expenditure and losses (26, 27). The 1992 consensus document for the nutritional management of CF (18) recommends that individualised gender- and age-specific energy requirements should be

determined, using basal metabolic rate, growth requirements and activity level as a basis (as for non-CF children) and taking into account pulmonary status and the degree of malabsorption.

In addition to inadequate energy, macro- and micronutrient deficiencies (due to decreased intake and gastrointestinal losses), lung infections and concurrent diseases can also affect growth and overall nutritional status. Nutrient deficiencies may inhibit growth either directly, or indirectly through compromising pulmonary function and immunity (24). Chronic lung infection can cause weight loss by increasing resting energy expenditure, decreasing protein synthesis and causing a large array of hormones and immune factors to be secreted, such as catecholamines and tumour necrosis factor-α (28). The study by Hull and Thomson (29) supports the possibility of a genetic contribution to the level of tumour necrosis factor-α in patients with CF. The metabolic effects of these stress hormones and immune factors have been difficult to determine but they are known to induce anorexia and increase energy expenditure (for example, by mobilising protein stores in muscle and fat stores in adipose tissue) (30). The presence of concurrent diseases (CF-related diabetes, liver disease and gastro-oesophageal reflux) also contributes to malnutrition through additional energy losses. The retrospective study by Lanng et al (31) revealed a decline in body weight, body mass index, FEV_1 and FVC, and an increase in pancreatic enzyme intake, in 3 to 40 year old pre-diabetic and control patients with CF for several years prior to diagnosis of diabetes mellitus.

Increased energy expenditure

Increased energy expenditure arises from a combination of lung infection and inflammation (through oxidant injury and inflammatory mediators), the increased work of breathing and coughing and the stimulation of metabolism by bronchodilator therapy (22, 32). A review of the literature on energy expenditure in CF reports that the increase in resting energy expenditure has been estimated to be between 9% and 30% in well individuals with CF, and that a cellular defect causing greater energy utilisation within the cell has been suggested, but also disputed, as the cause (33).

Studies investigating total daily energy expenditure in CF suggest that individuals compensate for raised resting energy expenditure by reducing their level of physical activity, such that those with moderate lung disease have comparable total daily energy expenditure to controls (34).

Maldigestion, malabsorption and other losses

Up to 90% of individuals with CF experience maldigestion, and hence malabsorption of nutrients due to exocrine pancreatic insufficiency, reduced bile salt pool and increased intestinal mucus (5, 24). Pancreatic insufficiency arises because abnormal cystic fibrosis transmembrane conductance regulator processing in the pancreas causes pancreatic enzyme secretions to be obstructed and bicarbonate secretions to be reduced. A small percentage (10% to 15%) of individuals with CF have variable degrees of pancreatic dysfunction but absorb nutrients normally and are classified as having pancreatic sufficiency (35). The incidence of pancreatic insufficiency and pancreatic sufficiency is thought to be linked to genotype, with a majority of delta F508 homozygotes having pancreatic insufficiency and worse prognosis (18).

Maldigestion in CF affects energy sources and protein. Growth may be limited by the subsequent energy and protein deficit and related increase in protein catabolism (36). Maldigestion of various micro-nutrients may also inhibit growth directly, or indirectly through the effects of specific deficiencies on pulmonary function and immunity (24). Gastrointestinal problems associated with maldigestion and malabsorption, such as abdominal pain, can affect nutritional status indirectly by decreasing appetite (37).

Long-term PERT is required by individuals with pancreatic insufficiency to improve macro- and micronutrient maldigestion. PERT aims to minimise the incidence of frequent, loose, bulky, foul-smelling, greasy stools, rectal prolapse, hypoalbuminaemia, oedema, anaemia and azotorrhea, which are commonly experienced before diagnosis. Refinements in pancreatic enzyme preparations over the past two decades have improved fat absorption such that, together with a high fat

diet, growth and nutritional status in CF have significantly improved. However, even though the currently used enteric-coated, encapsulated microspheres are much better than previous preparations, normal fat absorption does not appear to have been achieved by the majority of individuals on PERT, as indicated by several studies of subgroups of CF populations (11, 12, 38-40). Faecal losses continue to be high and may account for approximately 5% to 20% of gross energy intake in subjects with CF compared with 3% to 4 % in controls (32, 41). It is possible that protein digestion in individuals receiving PERT also remains sub-optimal as stool nitrogen output appears to be increased (9). However, this may be more a reflection of rapid colonic transit and antibiotic usage than an indication of the degree of protein digestion (9).

The effectiveness of PERT is thought to vary within and between individuals due to numerous factors related to the product (dose, potency, ratio of the various enzymes, microsphere size, shelf life, storage conditions) and to the host (residual pancreatic function, the action of lingual and gastric lipases, gastric hyperacidity, low duodenal pH, abnormal intestinal solubilisation of long-chain fatty acids, reduced mucosal absorption, liver disease, short bowel syndrome, type of diet and adherence). The variation in the degree of steatorrhea caused by these factors may be primarily related to genotype, as a majority of delta F508 homozygotes have more severe pancreatic insufficiency (18). Individuals with CF who have a high level of persistent malabsorption may need adjuvant agents, such as H_2-antagonists or proton-pump inhibitors, to improve the pH of the gastrointestinal environment and thereby increase the availability and effectiveness of pancreatic enzyme preparations. The achievement of optimal PERT in individuals with CF not only reduces maldigestion and malabsorption, but also ensures that the adverse effects of associated abdominal pain on appetite are minimised (37).

Small amounts of energy and nitrogen may also be lost from the body through the expectoration of sputum. Wootton et al (32) estimated this loss to be 1 to 5% of gross energy intake in five patients with CF during a period of hospitalisation related to an infectious exacerbation.

Inadequate intake

Pencharz and Durie (24) state that most growth problems in CF can be attributed to dietary inadequacy rather than to poor nutritional status being an inherent factor of the disease. This dietary inadequacy appears to be mostly due to unfavourable energy balance, although inadequacies of other nutrients could also contribute to poor growth. The extent to which nutrient inadequacies contribute is difficult to determine as the validity of measurements is uncertain and the specific requirements for CF are not known. For example, protein intakes of children with CF are commonly in excess of the RDI for age and gender (similar to children in the general population), but it is not known what level is sufficient to counter the effects of the disease as RDIs are developed for the general population, which mostly includes well children (42).

As previously mentioned, individuals with CF are thought to require between 120% and 150% of normal energy requirements for age and gender to compensate for elevated energy expenditure and losses (26, 43). Nutrition studies conducted in the 1980s indicate that in spite of increased energy needs, dietary intakes of individuals with CF averaged at only 80% or less of the requirements for the general population and few children ever achieved the elevated energy recommendation (44, 45). The observation of sub-optimal intakes in these studies is likely to be because they were reporting baseline measures of malnourished patients and that the data were collected prior to the change in policy for individuals with CF to consume a high fat diet rather than a low fat diet. However, more recent studies (12, 46-49) indicate that intakes continue to be well below the elevated energy recommendation, with mean levels ranging from 92% to 115% of the RDI.

The validity of studies reporting dietary intakes is limited by many methodological issues (dietary data collection method, representativeness of usual intake, type of food composition tables). In addition, the ability to draw conclusions about the effects of dietary intake on the course of pulmonary disease in CF, from a

comparison of nutrition studies is significantly limited by numerous factors, including:

- subject differences with regard to nutritional status, age, pulmonary function, disease severity, fat malabsorption, energy intake and energy needs

- variation in dietary recording methodology and nutrient databases

- variation between countries in nutrient recommendations, in definition and amount (eg. RDI for age and RDI for weight), and

- incomplete reporting of the data collected.

Although it is difficult to determine the exact extent of dietary inadequacies in CF, it is evident that many physiological, psychosocial and environmental factors cause intakes to be sub-optimal. Anorexia, dietary preferences, emotional problems, lack of knowledge about nutritional needs, poor adherence to a high fat diet and insufficient dietetic support can have an adverse influence on the oral intake of individuals (32). Factors associated with reduced appetite include acute and chronic respiratory infection, constipation, distal ileal obstruction syndrome, abdominal pain, fatigue and depression. A dislike for fatty foods, media pressure to eat a healthy low fat, low sugar diet, inappropriate concepts regarding body image, poor use of dietary supplements, behavioural feeding problems, parent-child meal-time interactions and a lack of financial resources can all contribute to reduced food intake (33). Interventions that minimise the effects of these factors are needed so that short periods of very low intake (with acute respiratory exacerbations) are not superimposed on prolonged poor intakes.

Concurrent diseases

Concurrent diseases may also contribute to the energy imbalance and poor nutritional status of those with CF, particularly prior to diagnosis and adequate treatment. Pancreatic impairment is often progressive and approximately 20% of older individuals develop CF-related diabetes. If untreated, or inadequately controlled, diabetes can contribute to energy deficits through glycosuria. In addition, liver disease may exacerbate the severity of malabsorption through inadequate bile acid

secretion. Other gastrointestinal complications (lactose intolerance, Crohn's disease, chronic abdominal pain, distal intestinal obstruction, gastro-oesophageal reflux and oesophagitis) may also make significant contributions to malnutrition in CF.

2.5 Assessing nutritional status

The 1992 USA consensus committee for the nutritional management of CF stated that there is no reason to accept nutritional failure and impaired growth (18). However, numerous studies suggest that progressive or episodic deterioration in growth and nutritional status continue to occur in those with CF (14). This may be because it is difficult to detect deterioration early due to the lack of generally accepted and validated markers of nutritional status in children. The heavy reliance on assessments of growth in children with CF may cause the extent of malnutrition to be largely underestimated. The usefulness of measures of height and weight is limited by:

- variation in the reference standards used to determine growth status
- differences in the criteria used to define growth and malnutrition (weight for height, weight for age, and expressing weight and height as percentiles, %IBW and z-scores), and
- variation in the malnutrition cut-off points between studies for each of these indices.

Borowitz (22) states that continuing to choose the %IBW index as the main measure for scoring systems in CF illustrates that there is uncertainty about how to determine nutritional status and the degree that malnutrition impacts on the course of the disease.

Since no single test provides an accurate measure of nutritional status, a variety of non-specific indicators from body composition, dietary and biochemical measurements are used to provide the basis for evaluation.

Body composition

The possibility that ongoing sub-normal growth in CF is due to low stores of fat and/or sub-optimal lean body mass (50, 51) suggests that there is a need for routine (quarterly to yearly) clinic assessments to include comprehensive measures of body composition. Monitoring of body composition may enable deteriorations in nutritional status to be detected early, and nutritional interventions to be implemented immediately, before inadequate weight gain, stunting in height and deterioration in pulmonary function occur (13, 52).

There are numerous methods available for assessing body composition, but most are invasive, expensive and not suitable for general use with children. The consensus report on nutritional assessment in CF by Ramsey et al (18) recommends that quarterly assessments of anthropometric measures, specifically mid-upper arm circumference and arm muscle area (derived from mid-arm skinfold and girth measurements), be compared to age- and gender-norms generated by Frisancho (53). Skinfold thickness measurements are thought to be useful as they indicate the level of subcutaneous fat and are considered to be an index of stored energy. Girth measurements are also useful for assessing nutritional status as they can be used to estimate muscularity. Lester et al (54) advocate that four anthropometric sites need to be measured in those with CF, namely triceps, biceps, subscapular and suprailiac skinfolds, in order to better represent overall body composition. Both the recommendations of Ramsey et al (18) and Lester et al (54) are in contrast to those of anthropometrists, who advise that numerous sites, from the upper and lower body, trunk and limbs, need to be measured so that variations in skin compressibility, skin thickness and tissue patterning with age, within and between individuals, are accounted for (55-57).

Dietary intake

There are numerous methods available for assessing dietary intakes, such as weighed intake records, dietary record diaries, duplicate portions, food frequency questionnaires (FFQs), 24 hour recalls and diet histories. The accuracy of assessments of dietary intake is limited by methodological problems. These problems are compounded in children due to the difficulties they may have in remembering their intake and their cognitive ability to complete dietary records (58). The technique chosen largely depends on the ability of the target group to comply with the method, the resources and time available, the study design and the purpose for collecting the information. There is minimal information in the literature regarding what tools are most advantageous for assessing the dietary intake of children.

Yearly 24-hour dietary recalls and 3 to 5 day prospective dietary records are the techniques most commonly used in the clinical setting to elicit pertinent information regarding an individual's nutritional and pancreatic enzyme pattern (18). This information is useful for assessing the adequacy of an individual's intake, determining where energy density may be increased, deciding how enzyme use may be optimised and for reviewing food-related behaviours (family dynamics, eating patterns and recent stressors) (59). Dietary records are also used in faecal fat balance studies (see biochemical assessment).

Questionnaires, such as a FFQ, are useful in the research setting, particularly if an aim is to compare the mean intake of nutrients between groups of subjects (60). A major advantage of FFQs is that they are less time consuming for subjects than other methods, thereby enhancing participation and completion rates (61). One specific disadvantage of FFQs that has been identified by the research conducted with children is that they can overestimate intakes for nutrients largely derived from fruit and vegetables (62, 63). The effects of FFQs on the estimated intakes of other nutrients by children is not known. The open-ended quantitative Geelong public health nutrition study FFQ, which was developed and validated for adults, appears to

be advantageous over other similar and available questionnaires as it is meal-based and has a comprehensive prompt list of over 300 foods (61, 64).

Biochemical assessment

Ramsey et al (18) recommend that electrolytes, acid-base status, complete blood count, serum albumin, retinol and alpha-tocopherol levels be assessed when an individual is diagnosed with CF, and if clinical deterioration is evident after diagnosis.

Evaluation of pancreatic function is also highly recommended, although no one technique is economical, simple to perform, non-invasive, specific, quantitative and reproducible (39). In the clinical setting, the faecal fat balance study is commonly used to indirectly assess pancreatic function and is considered the gold standard for assessing fat absorption. A 3 to 5 day dietary record and stool collection over the same period are used to assess total faecal fat output as a percentage of oral intake, with normal faecal fat excretion being <7% of the dietary fat intake of individuals who do not have CF (65). The faecal fat balance study is useful for determining the need for PERT, to investigate signs of malabsorption when on PERT and to assess the response to alterations in PERT (39).

2.6 Nutritional management of CF

Nutritional interventions are warranted in CF as improvements in nutritional status, through optimal dietary intake and PERT, could increase the number of individuals exposed to the survival advantage first demonstrated by Corey et al (3). The attainment of optimal nutritional status would ensure that individuals with CF have some energy in reserve for occasions when the maintenance of positive or zero energy balance is challenged by pulmonary disease (14). Optimal nutrition is also thought to enhance respiratory muscle strength and immunity, have long-term effects on the state of the lungs and ultimately improve prognosis (66) (page 1013). Thus, it

is important for nutritional interventions to be initiated early, when individuals with CF have mild or moderate pulmonary involvement; aggressive rehabilitation in those with forced expiratory volume in 1 second levels <40% is not always successful and may not be medically indicated in the terminally ill, unless the person is awaiting organ transplantation (21). Effective nutritional management of CF would ensure that both deterioration in nutritional status and the cause of negative energy balance are identified early so that nutritional interventions are prompt and targeted appropriately.

Pancreatic enzyme replacement therapy

The development of and refinements in PERT over the past two decades have enabled individuals with CF to tolerate much higher intakes of fat than when they were first available. Although there is little evidence to indicate how to determine optimum PERT dosage, current dosing guidelines suggest using dietary fat intake (67, 68) or body weight (69, 70) as a basis and then individualising the dose based on assessments of efficacy. The recently compiled Australian PERT Guidelines advocate fat-based dosing and also address numerous factors related to distribution, administration, storage, adherence and follow-up (67, 68). Minimum effective doses are recommended in order to decrease the risk of fibrosing colonopathy, which is characterised by severe sub-mucosal thickening of the bowel wall by mature fibrous tissue (71). Retrospective studies suggest that fibrosing colonopathy is associated with excessive doses and high strength pancreatic enzyme preparations and the type of enteric coating used on microspheres (67, 69, 70).

Nutritional requirements for cystic fibrosis

The nutritional requirements of individuals with CF are difficult to define due to the variable expression of the disease, clinical state and activity level. Although the RDIs for the general population are used as a benchmark for CF dietary recommendations, this practice is limited by the origins of RDIs and variations

between countries. RDIs are based on the general healthy population and are set at levels of intake that are considered to be adequate to meet the known nutritional needs of most healthy individuals so that deficiencies and diseases are prevented. Also, countries vary in both how they define nutrients and in the amounts recommended (72).

Following is a summary of the nutritional requirements of children with CF based on the USA consensus guidelines for the nutritional management of CF (18) and of the extensive reviews presented by Daniels (59) and McDonald (33). Nutritional status should be closely monitored in order to determine individualised needs, but the following recommendations are useful as a general guide.

Energy: Elevated energy expenditure and losses (due to malabsorption) in individuals with CF are thought to be compensated for by intakes between 120% and 150% of normal energy requirements for age and gender (26, 27). The CF nutritional consensus document details how to determine approximate energy requirements, using an age- and gender-specific estimated basal metabolic rate, growth requirements and activity level as a basis (as for non-CF children) and taking into account the patient's pulmonary status and degree of malabsorption (18). Inadequacies in weight gain or linear growth are suggested as the parameters to use to determine the need for a higher energy intake (18). However, this method of basing the need for more aggressive nutritional support on inadequacies in height and weight is inappropriate if the effects of malnutrition in CF are to be minimised. Wootton et al (32) recommend individual assessments of metabolic needs and appetite be conducted, but this is difficult as the equipment and expertise for indirect calorimetry is not available in most CF clinics. Alternatively, measures of body composition could be used to detect deterioration in nutritional status earlier than if changes in height and weight were relied upon (25).

Fat: In CF, fat intake should be as high as possible within the limits of individual tolerance and optimal PERT (59, 73), as a diet high in fat is less bulky and more likely to be achieved than a low fat, high energy diet of similar value. Also, the

energy cost of converting dietary fat to body fat is minimal compared with the conversion of dietary protein and carbohydrate to body fat. Nutrition guidelines have traditionally suggested that fat should provide 40% of the daily energy needs of individuals with CF if total energy intakes of more than 125% of the RDI are to be achieved (44, 74). However, Collins et al (12) found the positive association between fat, energy intake and growth to be stronger when fat was analysed as an absolute amount rather than as a percentage of energy intake. This led to the alternative and more practical recommendation for individuals more than 5 years of age to consume >100 g fat/day (12). In practice, parents can increase the fat content of the family's usual diet specifically for the child with CF by fortifying meals with margarine and cream, for example, and by providing additional nutritious high fat foods (cheese and milk-based desserts).

Protein: Currently there is no specific recommendation for protein. Ellis et al (42) report that protein intakes of individuals with CF are often well in excess of recommendations for the normal population which, for example, is 1 g/kg body weight for individuals between 1 and 18 years of age (72). Daniels (59) suggests that even if the protein requirements of children with CF are elevated, due to the excessive loss of nitrogen in the faeces and sputum and the possibility of altered protein metabolism, needs should be met if a high energy diet is attained and protein constitutes 15% of the energy intake (59).

Carbohydrate: Carbohydrate intake should be as high as possible, in conjunction with a high fat diet (59).

Vitamins: Little is known about the precise vitamin requirements of individuals with CF. Ongoing maldigestion and malabsorption in those receiving PERT is likely to affect the absorption of dietary fat-soluble vitamins more than water-soluble vitamins. Significant deficiencies of water-soluble vitamins have been rare in CF. In contrast, deficiencies of vitamins A, D, E and K have been repeatedly demonstrated (24). Many children with CF have been found to have low concentrations of vitamin

E and a neuropathy due to vitamin E deficiency has been widely reported in adults with CF (73).

Supplementation needs are not well defined as it is difficult to accurately assess vitamin status. Current recommendations include CF-specific supplementation of fat-soluble vitamins (A and E) and a normal dose of a water-soluble multi-vitamin-mineral mix. Caution with supplementation is necessary as individuals with CF, in comparison to normal controls, can have a 3.5-fold hepatic vitamin A reserve which could lead to hypervitaminosis if doses are excessive (33, 75). Vitamin K supplementation is reserved for those with liver disease and vitamin B_{12} administration is required by those who have had extensive ileal resection. Small amounts of vitamin D supplementation are necessary for individuals living in countries where there is limited sunshine (24, 73).

Minerals: CF is characterised by elevated sweat sodium and chloride concentrations. Salt depletion can occur due to sweating and in infancy due to low levels of sodium in breast milk and substitutes. Salt supplementation is particularly needed in hot weather, in conjunction with strenuous physical activity and in infancy to prevent acute hyponatremic dehydration (73). The possibility of chronic salt depletion contributing to anorexia should be taken into consideration when assessing the causes of inadequate oral intakes. Also, low iron or low zinc status can contribute to less than expected growth, levels should be monitored in individuals with CF and a trial of supplementation given if necessary (24).

Fibre: It has been suggested that the typically low fibre intakes of children with CF contribute to constipation and abdominal pain (76). High fibre diets are not universally recommended in CF, perhaps because the high satiety value of insoluble fibre may limit energy intake (59). The feasibility of individuals with CF increasing their intake of soluble fibre has not been reported.

Providing nutritional support

Enteral- and parenteral-feeding: Before the consensus paper on nutritional management in CF was produced by Ramsey et al (18), much of the literature focussed on approaches to artificial (oral, enteral and parenteral) supplementation in patients who failed to respond to routine dietetic counselling. Numerous studies (19, 20, 51, 77, 78) report the successful use of enteral- and parenteral-feeding (nasogastric, jejunostomy and total parenteral nutrition) to improve the nutritional status of malnourished individuals with CF. Markers of nutritional status (height, body fat, lean body mass, muscle mass, total body nitrogen, strength, development of secondary sexual characteristics and pulmonary function) were found to improve with supplemental feeding, particularly in those with only mild to moderate lung disease (18). During 1 to 2 years of follow-up of children with CF aged 3 to 13.2 years who underwent a 1 year course of semi-elemental, high-N enteral supplementation Shepherd et al (51) observed a decreased decline in pulmonary function status. This effect was attributed to the significant reduction in the number of pulmonary exacerbations and to the direct benefits of nutritional rehabilitation on declining pulmonary function, such as improved respiratory muscle strength and enhanced lung growth. Enteral-feeding and associated catch-up growth over 6 to 12 months were also found to reduce the high rates of both synthesis (which is energy expensive) and catabolism of whole body protein levels to values similar to those of healthy children, while maintaining normal rates of net deposition (51). However, the invasive nature of these methods and the problems experienced make them impractical as methods of early intervention. Problems that were encountered included cessation of tube feeding soon after initiation due to poor appetite, gastro-oesophageal reflux, vomiting, nasogastric-tube dislodgment, gastrostomy-tube blockage and site leakage, inconvenience with preparing and cleaning feeding sets and dissatisfaction with body image (33). Another disadvantage of enteral-feeding is that the benefits can be short-lived soon after supplementation ceases (79).

Oral route: Although there was a trend (non-significant) toward an improvement in weight gain with invasive methods of nutritional support in the meta-analysis of

treatment approaches to malnutrition in CF, Jelalian et al (80) suggest that oral supplementation and behavioural interventions with children and adolescents requiring nutritional rehabilitation can be as effective as enteral- and parenteral-feeding in improving weight gain. However, despite the marked improvements in palatability and presentation of oral supplements (fortified milk-based drinks and desserts and glucose polymers), their use is inconsistent and they are often inappropriately used as direct substitutes for normal food so that there is no overall increase in energy intake in the long-term (18, 33). The numerous disadvantages associated with oral supplements and enteral- and parenteral-feeding indicate the need for interventions that will enhance nutritional status by increasing oral intake.

The consensus document by Ramsey et al (18) supports the concept of anticipatory guidance, so as to optimise oral intakes and minimise deteriorations in health. Guidelines for the assessment of nutritional status and the provision of nutritional support are detailed in the report, including recommendations regarding frequency, such as measuring height and weight at 3 to 4 monthly routine CF follow-ups appointments. Ramsey et al (18) advocate that optimum nutritional management in individuals with CF includes the provision of anticipatory guidance by specialist CF dietitians and defined five response categories.

1. Routine management of all patients: high energy, high fat, high salt nutritional education and dietary counselling; and PERT and vitamin-mineral supplementation for those with pancreatic insufficiency.

2. Anticipatory guidance of patients at risk of developing energy imbalance, but who are maintaining adequate nutritional status: energy dense dietary education, close monitoring of dietary intake and behavioural management counselling.

3. Supportive intervention for patients with decreased weight velocity and/or slightly compromised nutritional status: anticipatory guidance plus oral supplements, such as fortified milk drinks and desserts and glucose polymers.

4. Rehabilitative care for patients whose nutritional status is compromised: overnight supplementation via an enteral tube.

5. Resuscitative and palliative care for patients whose nutritional status is significantly compromised: 24 hour continuous enteral-tube feeds and/or total parenteral nutrition, particularly in those awaiting organ transplantation.

It is possible that behavioural management training, together with nutrition education, could delay the need for oral and enteral-tube supplementation and decrease their use in CF by increasing oral intake and compliance with procedures (81). Jelalian et al (80) believe that early implementation of a behavioural approach to nutrition in CF, before dietary-related problems arise, could both prevent and minimise inadequate intakes and poor nutritional status.

2.7 Barriers to optimal oral intake

Complexity of medical treatment

Effective management of CF is influenced by the complexity of the treatment regimens. Medical advances have the potential to improve outcomes in CF, but the array of daily procedures (aerosol treatments, chest treatments, diet and pancreatic enzyme therapy), attendance at clinics and hospitalisations have a significant effect on family routine and priorities. Numerous resources are required from both the health-care system and the family in order to deal with the complications of the disease, which vary between patients in time of onset, severity and rapidity of progression. Over the long-term, CF treatments may strain the capabilities of the family to comply consistently with regimes, while also managing the normative tasks of family life (82). Consequently, no matter how technologically advanced medical treatments become they will only be as effective as adherence to their use (83).

There has been an emerging acknowledgment in the literature that a patient's choices deserve a greater degree of respect, such that there has been a shift in the literature from using the terms compliance to adherence to self-management (83). Children with CF and/or their parents need to be included in determining, monitoring and modifying therapies, in collaboration with health-care providers, so that perceptions,

motivation, skills and factors in the social environment are taken into account (84, 85). This may be particularly important with dietary intake and PERT in CF as the treatments are numerous and frequent, and have a significant effect on the family's lifestyle (83). Many of the different skills that are required for home treatments may not normally be present and need to be taught and mastered in order for children and their parents to effectively self-manage a chronic illness such as CF (86).

Knowledge

The acquisition of knowledge is important as it is one of the determinants of adherence to treatment (87, 88). Maternal CF-related nutritional knowledge has been found to significantly predict the level of dietary adherence in children with CF (89). Assessments of CF knowledge indicate that parents and children have significant misconceptions and gaps in knowledge about the disease (90), and about diet and PERT in particular (91). Despite the importance of a high energy, high fat, high salt nutritious diet and optimum PERT, children and parents are often unaware that diet is a formal treatment recommendation (88, 92).

Actual levels of CF nutrition and pancreatic enzyme knowledge and self-management practices are not known due to the lack of validated, comprehensive questionnaires. The assessment of the nutrition and pancreatic enzyme knowledge of children and adolescents by McCabe (91) is the only known published report that focuses on this aspect of CF care alone. Unfortunately, the questionnaire developed by McCabe (91) has limited application due to factors related to questionnaire design, namely, incompleteness of the topics addressed and non-establishment of readability, reliability and validity. Basketter et al (93) and Yuill et al (94) have presented the results of nutrition and pancreatic enzyme questionnaires in abstract form in conference proceedings only. From the limited information available it appears that the topics assessed by the questionnaires are not comprehensive, they assess an individual's practice rather than general knowledge and that readability, reliability and validity have not been established (93, 94). The questionnaires developed by Nolan et al (95) and Bartholomew et al (96) also have limited

application due to factors related to design, namely incompleteness of the topics addressed and assessment of other aspects of CF care, such as chest treatments and medication. Although attempts to assess CF-related nutrition knowledge have been limited, the process is justified as poor levels could make significant contributions to inadequate dietary intakes, absorption, growth and nutritional status in individuals with CF (95).

A family's nutrition knowledge may be overestimated on the basis of frequent clinic visits and familiarity with the dietitian, even though sessions are rushed and the advice given is mostly prescriptive. Intensive dietetic input can result in improvements in oral intake and weight gain of individuals with CF (45, 85, 97), but this level of service is not always available to all patients. Extensive patient reviews are often limited to one per year and much of the input is directed at the parents. In most situations there is insufficient time to equip families with the skills required to achieve nutritional changes (98). Nutritional services are also often limited by the far from ideal settings in which dietitians interview patients with CF. Counselling may occur in the busy waiting room area of the CF clinic or a separate dietetic office, that may be a considerable distance from the main clinic area where the rest of the CF team are working (73). These factors, and the possibility that there is limited reinforcement of the importance of nutrition by others in the CF team due to busy clinic schedules, may contribute to the dissociation by families of diet being a formal part of treatment for CF.

Traditionally, in most Australian CF Centres, families are counselled on the nutrition and PERT needs of the child at diagnosis and, thereafter, at outpatient clinics or during hospital admissions. Information is presented in verbal and written form and is mostly directed to parents. The retention of information during these sessions may be limited by a number of factors. Initially, much of the information presented at diagnosis is unlikely to be recalled as the emotional distress that parents experience at this time may be incompatible with optimum learning (99). Thereafter, at clinics and during hospitalisation, the provision of too much or fragments of information as problems arise may make it difficult for families to understand the relationships

between concepts (100). This is likely if several members of the CF team provide a family with numerous pieces of information on the same day or if the topic that is addressed is related to a crisis situation, such as inadequate growth.

Parents and children need continuous assessments and repeated learning opportunities in order to enhance dietary knowledge and be capable of making appropriate changes to the child's meals at all stages of development (81, 99, 101). It is important to provide a structured approach to health education, rather than only provide problem oriented counselling, as a comprehensive and accurate knowledge structure is needed to equip families with the ability to address specific issues or problems (100). The problem oriented approach was unsuccessful in a health education program that aimed to reduce the incidence of iron deficiency anaemia in infants (102). Teaching methods need to be imaginative and should accommodate the child's developmental stage and parents' different learning styles (99, 101). Suggested tools for children with CF and their parents include story and colouring booklets, nutrition games, reward stickers, certificates, computer assisted learning programs and supplement tasting sessions (73). A process of continuing education is needed in CF in order to dispel dietary misconceptions and myths, to promote a positive attitude towards elevated dietary requirements for the disease and to accommodate children's varying levels of comprehension according to their stage of development (59).

The imparting of knowledge in the best possible way is an important aspect for the nutritional management of CF, but it does not guarantee adherence to dietary recommendations (89). Parents and children may achieve a high level of knowledge (be aware of what children with CF should eat, know what foods are high in energy, fat and salt), but unless behavioural issues are addressed families may be unaware of the steps necessary to achieve the changes required. Parents may also have difficulty enlisting adherence in children with CF (103) and those with young children may be most in need of interventions to assist them in managing CF treatment-related difficulties (104).

Behaviour

Much of the CF nutrition literature focuses on mechanisms for providing a high energy, high fat diet with little acknowledgment of the problems faced by parents in getting their child with CF to consume the food and fluid provided. Physiological alterations (lung infections, hospitalisations and vomiting with coughing) may lead to anorexia. Subsequent food refusal can then adversely affect parent-child interactions and lead to behavioural feeding disorders (105). In comparison to non-CF families, parents of young children with CF have noticed more problem meal-time behaviours (excessively long meals, delay of eating by talking and spitting food out) (48, 104, 106). The more problems parents reported for their child with CF, the less energy the child consumed (106).

The quality of the infant-caregiver relationship, particularly feeding interactions, is thought to have a significant effect on the nutritional status of children with CF (107). The problem of inadequate oral intakes in CF is thought to be compounded by parental reactions to a child's refusal to eat. These parental reactions include increased anxiety, attending to non-eating more than to eating by forced feeding and increased attention through coaxing, offering less food, switching to preferred foods rather than risking food refusal or removal from the feeding situation, all of which may serve to maintain the child's avoidance (103).

Parental reactions are often fuelled by the emphasis placed by CF health-care providers on maximising children's dietary intake and weight gain (48). Parents may compensate for their anguish and guilt regarding CF by over indulgence and excessive leniency, making the process of establishing reasonable behavioural expectations and limits difficult (106). Studies of children with CF exhibiting problematic eating behaviours (103, 108-110) suggest that parents could benefit from participation in specialised behavioural management programs. The training provided could help parents use contingent praise and the setting of limits to reinforce appropriate eating behaviours in preference to strategies typically used

(coaxing, preparing a favourite meal) which may inadvertently reinforce non-eating behaviour (110).

2.8 Achieving nutrition-related behavioural change

Rapoport (111) suggests that the integration of nutrition information with motivational and behavioural components is more likely to achieve change than is direct persuasion and the enhancement of knowledge. Behavioural change is a highly complex process due to the interaction between psychological, cultural, environmental and behavioural factors. Behaviour therapy is based on the assumption that current behaviours have been learned largely from environmental events that preceded and followed the behaviour (98) and that change can be facilitated through strategies related to an individual's self-efficacy, locus of control, self-esteem, self-confidence, outcome expectancies, health belief and health value (112). In short, nutrition-related behavioural change is most likely to occur when nutrition education is integrated with behavioural psychology.

Self-efficacy plays a major role in explaining many health behaviours and may prove essential in planning nutrition interventions (113). Self-efficacy is defined as the degree to which people believe they are able or unable to overcome the difficulties inherent in performing a specific task in a particular situation (114), or in the case in question, the perceived ability to change dietary behaviour. Self-efficacy will affect which dietary aspects individuals with CF feel capable of changing, how much effort they will expend while trying to adopt a new behaviour and how long they will persist in the face of an obstacle (112).

The locus of control construct contends that people view the attainment of a particular behaviour as being either within their control (internals), where their action determines the outcome, or outside their control (externals) where reward is gained by forces other than one's self. Self-esteem refers to judgements of self-worth or the degree to which people like or dislike themselves. Self-confidence is the belief in one's own ability to succeed in one's efforts and achieve one's goals. Outcome

expectation is a person's judgement about whether changing their behaviour will produce a specific outcome. Health belief is the conviction of truth (or falsehood) of an association between two concepts and health value is the value placed on health outcomes (112).

Success with health education is more likely to occur when interventions using these constructs are directed by theories of health behaviours (115). Theories of health behaviour assume that individuals exist within environments where other people's thoughts, advice, examples, assistance and emotional support affect their own feelings, behaviours and health. The significant individuals and groups include family members, co-workers, peers, health professionals and other social entities who are similar to or influential for them (115).

When developing health education programs, one of the best models for addressing the psychosocial factors that determine behaviour is the social learning theory (115). The social learning theory has been successfully used in conjunction with a range of health-related behaviours, including smoking, exercise and diet (116). The benefits of using this over other theories of interpersonal influence (the theory of reasoned action, the health belief model, interpersonal communication, social network) is that the social learning theory comprehensively addresses both the psychosocial factors that determine health behaviours and the strategies to use to promote behaviour change (115). The social learning theory holds that behaviour, cognition and the environment all interact to determine each other and explain human behaviour. A basic premise of the social learning theory is that people learn not only through their own experiences, but also by observing the actions of others and the results of those actions (115). Table 2.2 summarises aspects regarding each of the social learning theory constructs (115).

Table 2.2

Social learning theory constructs

Concept	Definition	Application
Reciprocal determinism	Interaction between person & environment affects behaviour change	Involve others & work to change the environment
Behavioural capability	Knowledge & skills to influence behaviour	Provide information & training about action
Expectations	Beliefs about likely results of action	Incorporate information in advice about likely results of action
Self-efficacy	Confidence in ability to take action & persist in action	Point out strengths, use persuasion & encouragement, approach behaviour change in small steps
Observational learning	Beliefs based on observing others like self &/or visible physical results	Point out others' experience, physical changes, identify role models to emulate
Reinforcement	Responses to a person's behaviour that increase or decrease the chances of recurrence	Provide incentives, rewards, praise, encourage self-reward, decrease possibility of negative responses that deter positive changes

Adapted from Glanz and Rimer (115)

Experts recommend that behavioural constructs should be used simultaneously to explain and address complex food and diet-related behaviours as their effects are markedly reduced when used alone (112). For example, AbuSabha and Achterberg (112) report that not all dietary behaviour studies have observed a relationship with locus of control, probably because the ability of this construct to predict health behaviours is more significant when used in conjunction with other constructs, such as outcome expectancy and self-efficacy.

2.9 Features of an ideal health education program for children

The challenge in health promotion is to translate a theory-based model into a program that includes a series of learning activities which will influence behaviour change (96). An effective intervention is one that provides information to increase awareness of health needs; facilitates behavioural change, mastery of skills, problem solving and open communication; and fosters self-efficacy and optimal functioning (117). Optimal functioning in the family is imperative as parents need to manage many aspects of treatment depending on the child's cognitive development and motor ability (101).

Following is a list of program strategies which numerous studies and reviews (88, 96, 118-123) recommend be used in health education programs for children. The strategies are based on social learning theory constructs and are useful for teaching children what behaviours are required and how to develop the behaviours.

1. Provide clear information in developmentally appropriate ways.

2. Encourage parental reinforcement of the advice provided & the appropriate behaviours exhibited.

3. Involve the family unit.

4. Establish concrete behavioural goals.

5. Allow the child to determine and participate in appropriate aspects of treatment.

6. Develop mastery over treatment skills.

7. Introduce record keeping in order for the child to self-monitor adherence and progress with behavioural goals and to assume more self-responsibility and awareness. Records are also a useful basis for feedback from the health-care provider.

8. Provide interim follow-up through telephone calls and checking of self-monitoring records.

9. Implement a structured reward system, in which the child chooses the rewards.

10. Develop problem-solving skills.

11. Provide opportunities for actual experience and participation.

Some centres in Australia conduct group education programs for children, separate from parents. The use of the group format is particularly encouraged in behavioural-based health education as a means of providing opportunities for observing others who are in a similar situation and for receiving support and social reinforcement of desirable behaviours (101). Group sessions are also useful for developing open communication within the family and for enhancing relationships between health-care providers and families (101). Levels of self-efficacy can also be enhanced through the group format as parents and children are encouraged about their capabilites by other families and the health-care provider. There are also several advantages of the group format for the health-care provider, namely that sessions can be cost and time effective and can provide an opportunity to gather additional information for assessing parental understanding and coping mechanisms (81, 101).

However, group learning may not be suitable in all situations. Some parents may not be available to participate in several weekly or fortnightly sessions and the motivation of others may decrease with time (92, 122). When Bartholomew et al (92) piloted a cystic fibrosis program, only 23% of families attended at least one of four parts of the group oriented program. Parents were pleased the education programs were available but could not devote the time to attend. Based on these findings, Bartholomew et al (96) wrote a home-based program in order to reach the majority of their population of families who had a child with CF. Bartholomew et al (92) concluded that the design of health education programs for individuals with CF needs to ensure that families do not have to expend extra effort to access educational sessions.

Support for home-based programs was found in the research findings of Perry et al (122) which assessed health-related behaviour change, associated with heart disease prevention, within the family context. The importance of parents' influence on children's behaviours was acknowledged, such that parents were asked to contribute to the development of the Hearty Heart Home Team general nutrition program for primary school-aged children. During the planning stage, parents stated that they would prefer a home-based program that included printed materials or activities,

for individuals with CF have been reported in the literature and all were based on social learning theory constructs. The treatment approaches by Stark et al (109, 110, 125) focussed on parent training to manage children's problematic meal-time behaviour; Luder (85) developed counselling strategies to nurture nutritional self-management skills in individuals with CF; and Bartholomew et al (92, 96, 113) developed a home-based print curriculum known as the Family Education Program. Following is an assessment of each of these three programs with regard to the use of social learning theory constructs (as summarised in Table 2.2) and the features recommended for an ideal health education program for children (as listed in Section 2.9).

Behaviour management training

Stark et al (109, 110, 125) report a series of interventions that targeted mal-adaptive parent-child meal-time interactions. The interventions consisted of group sessions, that were conducted separately for parents and children with CF aged between 3 and 12 years. Group sessions were used as the format to provide nutritional information, positive reinforcement of appropriate eating from the health-care provider and parent training on behavioural child management skills to modify problematic meal-time interactions. The group format also provided an opportunity for observational learning and reinforcement as children were provided with opportunities to practise eating new foods with others. The child management skills addressed during the program included differential attention, contingency management and implementation of meal-time rules and consequences. Behavioural capability and self-efficacy were nurtured through the provision of specific guidance regarding incremental dietary changes to make each week, such that the energy intake of children increased by 1 000 to 3 000 kJ/day, pre- to post-treatment (109, 110, 125). The reports failed to indicate what the changes in energy intake meant for individuals in terms of RDIs. Significant changes in height and weight were observed during treatment and the year following the interventions, but not in pulmonary function. Only one meal and one set of parenting behaviours were presented per group session and the information obtained built upon the knowledge and skills learned in the

previous session (125). The children were rewarded for achieving daily energy goals at each meal and for following meal-time rules. One aspect that is not clear about these interventions is the comprehensiveness of the nutritional information provided. It appears that families were simply advised what to increase without the provision of structured information about all nutritional aspects related to CF (energy, fat, salt, malabsorption and PERT). Also, participation rates in the clinic-wide implementation of a program which is entirely group-based could decrease with time.

The studies by Stark et al (109, 110, 125) have provided extensive insight into the behaviour management needs of parents of children with CF. Even though most of the interventions were designed for a small number of mildly malnourished subjects (n=3 to 5), with pre-existing, problematic eating behaviours, the techniques used to train parents could be modified for use in a prevention program. The aim of a clinic-wide prevention program could be to minimise inappropriate parental reactions to food refusal and to enhance positive reinforcement of desirable food behaviours in children. A program with this focus could be time and cost effective through the prevention of problematic meal-time behaviours and subsequent poor nutritional status.

Development of self-management skills through counselling

Luder (85) used social learning theory constructs to train individuals with CF aged 4 to 29 years (mean age 13.5 years), irrespective of the severity of the disease, in nutrition-related self-management skills. Patients identified and analysed their eating problems with the dietitian, set goals and monitored adherence to their plans. Self-management skills were developed during an average of five counselling sessions, over one to two years, until the desired goal of a mean energy intake of 115% RDI was achieved and weight status improved (85). Luder (85) reports that the mean intake of energy increased from 93.6% of the recommended dietary allowance to 125.8%, mean body mass index increased from 16.9% to 18.8% and pulmonary function remained unchanged during the intervention. The mean increase in energy intake was clinically significant as the intake of more of the subjects' was within the

elevated range recommended for individuals with CF (120% to 150% of the RDI). It is important to note that the change in body mass index may not have been clinically significant as it may simply reflect the variation that occurs with age in children (126). Luder (85) states that the development of self-management skills regarding how to increase energy intake and adjust pancreatic enzyme doses to control steatorrhea helped convince patients that they could manage their own treatment regimen effectively.

During the counselling sessions patients were guided to set short-term goals, such as consuming a milkshake every evening. The use of incremental steps to achieve the larger goal of a higher energy intake and subsequent weight gain aimed to enhance patient's self-efficacy and self-motivation. The dietitian also used verbal persuasion and encouragement to enhance patient's belief that they could achieve the dietary changes required. Problem-solving abilities were developed by guiding patients to make changes in the diet plan whenever necessary. Patients were encouraged to actively participate in the intervention and to self-evaluate rather than simply comply. However, self-evaluation was mostly based on information obtained from 24-hour dietary recalls and height and weight measures. Feedback from the dietitian based on the dietary recall would have been helpful in the learning process, but may not have been frequent enough to act as a constant motivational factor. Also, there is the possibility that the dietary recalls may not have been accurate as patients may overestimate intake toward expected levels. Daily recording of adherence to short-term goals would have been a more beneficial source of reinforcement. For example, monitoring whether or not a milkshake was consumed each evening as planned and rewarding oneself for progress with goals each week could be an important reinforcement strategy. The use of tangible (physical) rewards is another method of reinforcement that helps motivate people to continue participating in a program and to reinforce behaviour change in children (122, 124). The use of extrinsic reinforcement was not discussed in the report by Luder (85). This may be because the benefits of rewards in a behavioural change program were not recognised as the participants were not all children.

Luder (85) states that the nutrition guidelines provided during the counselling sessions consisted of general information about eating a variety of foods and PERT. The possible lack of structured, comprehensive information may have been because the program goals were limited to two aspects, energy intake and PERT dosage. Additional information, specific to the performance of self-management behaviours regarding fat, salt, malabsorption and PERT distribution, administration and storage, would also need to be addressed in a nutrition intervention program aimed at assisting families in being capable of meeting the extensive treatment needs of children with CF.

Also, more comprehensive nutritional information and a greater range of reinforcement techniques, together with other strategies in addition to the one-on-one sessions between the patient and dietitian, would need to be included in order for the counselling approach developed by Luder (85) to be suitable as an intervention program for children. The addition of a group format would be advantageous as it would provide opportunities for reinforcement through social support or modelling by other patients. Observational learning, in settings other than just during clinic visits, would be useful as numerous distractions in the clinic waiting area and concerns with other aspects of CF treatment would limit the learning ability of families. The time intensiveness for the health-care provider of the counselling sessions advocated by Luder (85) would be a limiting factor and could affect the successful application of this approach in the paediatric clinical setting.

The CF Family Education Program

The CF Family Education Program developed by Bartholomew et al (92, 96, 113) was a home-based print curriculum that consisted of four modules (nutrition and malabsorption, respiratory, coping and communication) with separate material for four age groups (early childhood, middle childhood, adolescence and parents). Bartholomew et al (92) recognised the advantages of using the group format in educational programs for families who have a child with a chronic illness during the initial planning stage, but parents who participated in the pilot program indicated that

they did not want to attend sessions at the CF centre outside of usual clinic attendance. Based on this feedback, the curriculum that was developed for the Family Education Program was entirely home-based. The concept of home-based nutrition programs for children, in which parents are involved, is supported by research regarding general nutrition programs for school-aged children (122). However, non-inclusion of the group format in the CF Family Education Program may have limited the development of self-management skills of some families who benefit greatly from observational learning and social reinforcement available in this type of setting.

The aim of the CF Family Education Program was to maximise health status through the mastery of self-management skills based on three social learning theory constructs, namely behavioural capability, self-efficacy and outcome expectations. Close examination of the program indicated that observational learning through modelling and social reinforcement were the only social learning theory constructs used in the nutrition module for children (96). The nutrition module for children depicted two characters modelling self-management skills and co-operation with tasks, such as eating enough and taking enzymes. Although the reason for limiting the information in this module is not stated, it may be related to the authors' caution in imposing responsibility for self-management tasks in children too early (96). However, the lack of comprehensive information may have prevented children from knowing what steps they could take to achieve desirable nutrition goals. Non-inclusion of developmentally appropriate information, and the omission of guidance through a series of incremental steps towards optimum dietary intakes, is surprising given that the authors acknowledged the importance of these social learning theory-based strategies for enhancing behavioural capability and self-efficacy (113).

The lack of comprehensive information and specific meal- and snack-based behavioural goals in the children's nutrition module of the Family Education Program may have alternatively been an oversight as a consequence of the apparent non-inclusion of a dietitian/nutritionist in either the multidisciplinary research team or expert panel validating the performance behaviours for the program (86). The

nutrition-related objectives of the program (for all four age groups) appear to be very general and restricted to: the administration of pancreatic enzymes, monitoring of weight, food intake and malabsorption and maintenance of adequate dietary intake. Other very important nutritional aspects should also have been included because dietary and pancreatic enzyme regimens for individuals with CF require a great deal of planning and extensive problem-solving skills as they interfere with activities of daily living more so than a list of prescribed medications (88).

As previously mentioned, another aspect of concern about the effectiveness of the Family Education Program for children was that reinforcement of the fostered behaviours was restricted to social reinforcement from program participants, health-care providers and the family. Interestingly, primary school-aged children participating in the program also received certificates for skill completion, indicating a recognition of the need for extrinsic rewards to reinforce behaviour change in children. The authors stated that tangible rewards for behavioural achievement were not included in the program because of their varying impact among patients and families and possible negative effects from extrinsically rewarding an action that must ultimately become intrinsically reinforcing (96). However, behavioural experts and studies assessing the motivational needs of children involved in health-related behavioural change programs (122, 123) stress the need for regular reinforcement through tangible rewards, phone calls and record keeping. The extrinsic reinforcers can be phased out once the behaviours have been adopted (124).

Bartholomew et al (127) recently reported that the CF Family Education Program resulted in significant improvements in a range of parameters, including children's knowledge and self-efficacy and carers' knowledge, self-efficacy and self-management skills. However, the questionnaires, which contained approximately 50 questions for each age group (children, adolescence and parents), assessed aspects regarding all four modules (nutrition and malabsorption, respiratory, coping and communication) at the same time. Details were not provided regarding how many questions assessed nutrition and PERT or if the improvements that occurred were with these topics. Given that the nutrition module for children did not include

sufficient strategies for achieving health-related behavioural change (Section 2.9) it is unlikely that significant improvements in dietary and PERT knowledge, self-efficacy and self-management were observed.

2.11 Aim of this research

This review of the literature highlighted the need for nutrition interventions which address the psychosocial issues affecting the oral intakes of children with CF and aspects regarding PERT. The literature suggests that prevention programs which incorporate strategies for behavioural change are more effective than those that simply provide nutrition information. Numerous deficiencies with regard to the features recommended for an ideal program for children were identified when three CF nutrition programs (85, 92, 96, 109, 110, 113, 125) were closely examined. The conclusion to be drawn from the literature was that there is a need for a behavioural-based nutrition prevention program specifically for children with CF to be developed. Such a program should incorporate behaviour management strategies together with nutrition information in order for the dietary intake and PERT of children with CF to be maximised, so that nutritional status is optimised, quality of life is improved and survival is enhanced. This review of the literature also indicated the need for validated, comprehensive CF nutrition and pancreatic enzyme knowledge and self-management questionnaires to be developed for assessing children with CF and their carers.

The focus of this thesis is the development and evaluation of the *Go and Grow with CF* nutrition and behaviour intervention program. Chapter 3 describes the development of the program and the process of developing and validating knowledge and self-management questionnaires to assess *Go and Grow with CF*. Chapters 4 and 5 detail the methodology and results obtained of the process evaluation of *Go and Grow with CF* and of the clinical trial designed to evaluate the effects of the pilot intervention program on knowledge, self-management, behaviour, dietary intake and body composition. Chapter 6 describes the clinical implementation of the revised program and an assessment of fat-based pancreatic enzyme dosing on absorption.

3. CHAPTER THREE: PROGRAM & QUESTIONNAIRE DEVELOPMENT

The lack of behavioural-based CF nutrition programs that have been specifically designed for children led to the development of *Go and Grow with CF*. The *Go and Grow with CF* education and behaviour change program differs markedly from the three previously mentioned nutrition programs (85, 92, 96, 109, 110, 113, 125), particularly in the application of two social learning theory concepts. *Go and Grow with CF* provides comprehensive information targeted for children of primary school age (behavioural capability) and a greater range and a more structured approach to rewards (reinforcement). Other specific features that were incorporated into the program to facilitate behavioural change included the combinaton of group (observational learning) and home-based learning (reinforcement) and a system of small incremental steps to facilitate behaviour change (self-efficacy). Table 2.2 contains a description of these concepts and other social learning theory constructs that were used as a basis for developing *Go and Grow with CF*.

The aim of this chapter is to describe in detail the development of and the features unique to the *Go and Grow with CF* program. The chapter also includes details regarding the development and validation of knowledge and self-management questionnaires to assess the effects of the program.

3.1 Needs assessment

The CF dietetic service provided through Princess Margaret Hospital is based on the CF nutritional assessment and management consensus document by Ramsey et al (18) (as outlined in the sub-section regarding the provision of nutritional support in Section 2.6). Families are counselled on the nutrition and PERT needs of the child at diagnosis and, thereafter, at outpatient clinics or during hospital admissions. Information is presented in verbal and written form and is mostly directed to parents. The retention of information during these sessions may be limited by a number of

factors including the emotional distress that parents experience at diagnosis and the provision of fragmented information as problems arise at busy clinics and during hospitalisation. Dietetic counselling and education most often occurs in the waiting area or in the nutrition department, which is located some distance away, due to a lack of space in the outpatient clinic.

Informal assessment during clinic reviews of nutrition and pancreatic enzyme knowledge and practice indicated that primary school-aged children did not know standard information and/or did not implement recommendations provided. A review of the literature and discussions with dietitians with CF expertise in Australia highlighted the lack of appropriate programs to address the nutritional needs of children with CF.

Focus groups were conducted on two occasions to assess the need for a change in the approach to CF nutrition and PERT education at Princess Margaret Hospital. The focus groups were scheduled for an hour before two CF clinic sessions commenced as Bartholomew et al (92) suggest that parents often cannot devote the extra effort to attend additional CF-related activities. Children aged 7 to 12 years and carers of 0 to 12 year olds who were scheduled to attend these CF clinics were invited by phone to participate in a nutrition-related focus group discussion. Nonetheless, the number of families involved in the focus groups was limited by the non-willingness of carers to arrive at the hospital at an earlier time, spend extra time at the hospital and their apparent lack of interest in nutritional aspects regarding CF. The two focus groups involved nine and six carers of 2 to 11 year olds, respectively, and four and six children aged 7 to 11 years, respectively, who met separately from their carers. Health professionals not involved in the CF clinic facilitated and recorded the discussions.

Assessment of the focus group discussions identified a number of issues:

1. Carers were interested in learning more about nutrition and enzymes, and agreed that a new approach to nutrition education would be worthwhile.

2. Most carers enjoyed meeting with other families to discuss aspects of caring for a child with CF, but their interest in attending extra sessions at the hospital was limited.

3. Carers preferred teaching sessions to be held on the same day as clinic sessions to minimise the number of trips to the hospital, particularly for rural families. There was a preference for the sessions for children to be conducted during school holidays.

4. Carers of 10 to 12 year old children suggested children should be involved in an education program from a young age.

5. Carers thought most interest in a new nutrition program would come from families whose child had recently been diagnosed with CF, such as in the last four years.

6. Carers identified that child-care facilities for siblings would be an important factor in maximising participation in group sessions.

The focus group sessions with children did not reveal any information that influenced the development and implementation of a new approach to nutrition education. Many of the children did not answer the questions that were asked, apart from what food they like to eat. The interviewers reported that the children appeared unconcerned and self-assured about CF nutritional issues and appeared to not want to discuss problems associated with eating. The majority of children appeared to the interviewers to be ambivalent toward the notion of a new approach to nutrition education.

The focus groups, together with the review of the literature, provided sufficient support for a new approach to nutrition and PERT education for children with CF at Princess Margaret Hospital. Even though carers in the focus groups thought that most interest in a new nutrition program would come from families whose child had recently been diagnosed with CF, a program for children with CF aged 6 to 11 years and carers of 2 to 11 year olds was developed. Parents involved in the focus groups were unaware that the nutritional needs of families with a child newly diagnosed with CF were being attended to through an annual workshop, which had been launched in

the same year that the *Go and Grow with CF* program was conceived. The subsequent availability of newly designed nutrition information for parents of children with CF (128) also highlighted the need for material specifically directed at children with CF.

3.2 Objectives of the intervention program

Validation of CF performance objectives (behaviours that patients and their families need to do in order to adequately manage the disease) by Bartholomew et al (86) suggested that the provision of specific teaching of families regarding self-management is often overlooked. For example, most centres expect individuals with CF to maintain an adequate intake of food. However, less importance is often placed on the provision of guidance regarding how to achieve a specific related behaviour, such as supplementing intake with energy-dense dietary sources (86).

It is difficult to determine what is an appropriate level of nutrition and pancreatic enzyme knowledge for children with CF and their families due to the lack of assessments using validated tools (Section 2.7, knowledge). Healthy, non-CF, children are expected to know the difference between foods that they should eat every day and foods that should only be consumed occasionally (122). Therefore, it may be reasonable to expect children with CF to know at least this type of information in relation to CF.

The review of the literature and health education programs, and the suggestions of dietitians with CF expertise in Australian paediatric centres, were used as a basis for developing the objectives of the intervention program. The nutrition and behavioural objectives of the proposed intervention program for children with CF aged 6 to 11 years and carers of 2 to 11 year olds were as follows:

Enzymes

- appraise optimum administration
- determine distribution according to the fat content of meals and snacks
- describe digestion and the role of pancreatic enzyme supplements
- identify signs of malabsorption, and
- determine how to monitor malabsorption.

Dietary

- identify food and drink high in energy, fat, salt and/or nutrient density
- determine how to increase the daily intake of energy, fat, salt and nutrient density
- discover why children with CF need an energy, fat, salt and nutrient dense diet
- discuss the consequences of inadequate energy, and
- identify the signs of poor growth.

Behaviour

- appraise enzyme problems (eg. not remembering to take enzymes, refusal, inappropriate administration, malabsorption)
- appraise meal and snack problems (eg. inadequate intake, refusal)
- discover problem solving strategies
- evaluate, formulate and practise ways of dealing with meal-related problems
- translate nutritional needs into daily meal and snack plans, and
- discover how to reinforce learning and behavioural change.

3.3 The behavioural change model

I developed the *Go and Grow with CF* program in consultation with a psychologist. The specific aim of the program was to facilitate families in the process of translating nutrition and PERT knowledge into behaviour and action. The constructs on which *Go and Grow with CF* was based were those of the social learning theory model of behavioural change (115, 129) (Table 2.2), particularly behavioural capability, self-efficacy and reinforcement. The recommendations of Perry et al (122) (Section 2.9) were also used as a basis for developing the practical behaviour change activities.

Figure 3.1 illustrates the design of the program which was developed for primary school-aged children (aged 6 to 11 years), and carers of 2 to 11 year olds. A combination of group sessions and home-based learning was used, with the main focus being learning in the home environment. Comprehensive nutrition and PERT information was included in the home-based course to equip families with the knowledge and skills required to be capable of achieving desirable behaviours (behavioural capability). The home-based course then guided families through six incremental steps to achieve desirable behaviours regarding the seven nutrition and PERT topics (self-efficacy) (Figure 3.1). The steps were:

1. *Plan:* The family specified a change to food selections or enzyme therapy based on the week's topic, and planned a reward to enjoy at the end of the week for participation and/or progress.

2. *Do It:* The change was tried for a week and recorded by daily monitoring.

3. *Feedback:* The family reviewed the impact of the change, modified the plan if necessary, and enjoyed the reward (reinforcement).

4. *Practise:* The revised plan was implemented.

5. *Keep Going:* The change continued, with rewards at regular intervals to maintain participation.

6. *Get There:* The family incorporated the new behaviour as part of their daily lives.

Figure 3.1

The *Go and Grow with CF* program structure

PROCESS   SOCIAL LEARNING THEORY

Introductory workshops for children and carers

$\Rightarrow$

A home-based course which covers seven $\Downarrow$ Behavioural capability
topics and guides children and carers through   (comprehensive information)
six steps to achieve behaviour change

   1. Plan $\Downarrow$ Self-efficacy
   2. Do It   (small incremental steps)
   3. Feedback
   4. Practise $\Downarrow$ Reinforcement
   5. Keep Going   (self-monitoring,
   6. Get There   rewards, newsletters,
                                                                phone calls, workshops)

$\Rightarrow$

Concluding workshops for children and carers

Where appropriate, behaviours were built upon during subsequent weeks. For example, at the beginning of the energy and fat topic, children were guided to plan a high energy, high fat breakfast every day for a week. The following week the focus was high energy, high fat morning snacks. The children were challenged to consume these more often than they had consumed their planned breakfasts the week before. A third week was spent on high energy, high fat supper snacks. Over progressive weeks the children were encouraged to continue with the behaviours already developed. The purpose of planning daily meals or snacks, and challenging children to consume these more often each week, was to help them reach the long-term goal of a high energy, high fat intake every day.

Reinforcement was also considered to be a key social learning theory concept to include in *Go and Grow with CF*. As achievement of a high energy, high fat, high salt, nutritious diet and appropriate PERT requires a large degree of behavioural change, carer involvement was essential in order to provide social reinforcement. Weekly rewards for participation with daily plans was also a source of reinforcement (external). Behavioural experts suggest children are more likely to repeat appropriate behaviours, and have positive expectations about a program, if they value the rewards (123, 124). Therefore, carers and children were guided to prepare the reward list together to ensure that the activities and items were valued by the child.

Another source of reinforcement in *Go and Grow with CF* was the task of recording adherence to daily plans. This activity of self-monitoring provided children with immediate feedback about their ability to change, and was a basis for the dietitian to provide external reinforcement when completed worksheets were reviewed once per month. Additional external reinforcement for families during the home-based course came in the form of fortnightly newsletters and monthly telephone contact between the carer and dietitian. Parents who participated in the general nutrition program for primary school-aged children by Perry et al (122) (Section 2.9) indicated that they preferred these strategies over evening information sessions as methods for parent education.

3.4 The intervention program

As shown in Figure 3.1, the *Go and Grow with CF* program began with separate, concurrent introductory workshops for primary school-aged children and carers of 2 to 11 year olds to introduce nutrition and PERT learning objectives, explain the home-based course and to motivate families to participate and complete the program. The workshops were followed by the home-based course, which consisted of printed materials and ten weekly activities, for children and carers to complete together. The seven topics covered in the home-based course were pancreatic enzyme supplementation, increasing fat, salt and micronutrient intake, malabsorption, growth and the importance of snacks. Finally, separate, concurrent concluding workshops for children and carers were held to review their learning and celebrate participation and progress throughout the program.

The introductory workshops were conducted during a set of school holidays, the home-based course was completed during the school term, and the concluding workshops were held during the following school holiday. The workshops for carers and children were held in informal settings at the hospital and were conducted by the CF clinic dietitian and other dietitians who participated on a voluntary basis. The written material for the home-based course was presented to families at the end of the children's introductory workshop. Families who did not attend the introductory workshop received the home-based course by post and implementation advice by telephone.

The home-based course was designed to be complete in itself in meeting all the nutrition, enzyme and behaviour objectives of the program (Section 3.2). Appendix 3.1 lists the contents of the revised program and contains the PERT module of the revised home-based course.

During the home-based course, the child and carer met once each week:

- to review their participation and/or progress with the previous week's topic

- to discuss strategies to improve or continue practising their new skill

- to decide when to enjoy their reward for participation and/or progress the previous week

- to read information and complete the activities for the next topic

- to plan a reward for participation and/or progress for the following week, and

- to plan what they would do each day of the coming week relevant to the topic.

Each day of the following week, the child or carer recorded if the planned changes were achieved. The activities in the home-based course took families approximately one hour per week to complete.

3.5 Development & validation of knowledge & self-management questionnaires

During the planning phase of this research, the lack of appropriate and validated instruments for measuring the nutrition and pancreatic enzyme knowledge and self-management practices of children with CF and their parents was identified. The few instruments which have been used to assess nutrition and pancreatic enzyme knowledge and self-management skills were considered to have limited application due to factors discussed in Section 2.7 (knowledge). Because suitable questionnaires would be useful in both the research and clinical setting, and were necessary for this research project, a study was undertaken to develop and validate nutrition and pancreatic enzyme knowledge and self-management questionnaires for use with children who have CF and their carers.

Methods

Questionnaire design

Separate, but similar, questionnaires were developed for children and carers in consultation with a research psychologist, based on the learning objectives of the *Go and Grow with CF* intervention program as listed in Section 3.2. The questionnaires

were designed to contain at least two brief questions assessing each aspect of the learning objectives, where this was possible. The knowledge and self-management questionnaires were designed to be administered as an interview, by telephone with carers, and face-to-face with children during a CF clinic visit, in order to minimise the impact of the respondents' variable reading abilities. Several of the children's knowledge questions were illustrated with large colourful food pictures to aid the child's concentration and comprehension. Care was taken to ensure the size and colouring of the food in the pictures for individual questions were equal to avoid bias.

Knowledge questions were mostly closed, in that respondents selected one of two alternative answers or "don't know". The alternative response of "don't know" was provided to discourage guessing.

The self-management questionnaire consisted mostly of scenarios about which the respondents were initially asked an open question in order to assess the behavioural components of the learning objectives (Section 3.2). For example, "If your child had signs of more than usual malabsorption what would you do?" Several appropriate and inappropriate responses were possible for each question. When respondents indicated they had finished answering the open question, prompts were read out from a list of possible answers not yet stated in order to assess if respondents knew a range of possible correct responses. An example of a prompt is, "If your child had signs of more than usual malabsorption would you: review what he/she has been eating; review the number of enzymes; increase the number of enzyme capsules each day, etc?" Yes, no or don't know responses were required for each item on the list. Prompts were not given with the few questions that asked the respondent to recommend the number of pancreatic enzymes that should be taken by the character in the scenario.

The children's self-management questionnaire also included questions assessing signs of possible malabsorption and communication with their carer about

management. The carers' self-management questionnaire also included questions to assess the age at which they expected children to manage their own PERT.

The questionnaires were reviewed by colleagues with a wide range of expertise, such as dietitians working in a variety of clinical areas, psychologists and medical researchers. Feedback was used to redraft the questionnaires in preparation for pre-testing.

Pre-testing

The questionnaires were pre-tested with six children with CF and their carers. The number of children selected for this process was limited by the small CF clinic population, the necessity for the respondents to resemble the target population (in age and pancreatic status), but be otherwise ineligible for the clinical trial and the willingness of families to be involved. Respondents were interviewed by health professionals not involved in developing the questionnaires.

Comments from both interviewees and interviewers were used to revise the questionnaires and the interview process in preparation for piloting. The type of feedback requested included information regarding question difficulty, misinterpretation, respondent interest and attention, flow of the questionnaire, order of the questions and timing.

Pre-testing also provided an opportunity to pilot other instruments to be used in the clinical trial (Chapter 4 & 5), namely the behaviour and dietary questionnaires; the household record form; and the anthropometry, lung function, illness and activity recording sheets.

Piloting

The children's pilot knowledge and self-management questionnaires contained 38 and 14 questions, respectively. The carers' pilot knowledge and self-management questionnaires contained 53 and 17 questions, respectively.

The questionnaires were piloted with 20 children with CF aged 6 to 11 years, and 20 carers of 2 to 11 year olds. Children and carers involved in piloting were a combination of those excluded from the clinical trial and those to be enrolled. It was necessary to include children and carers in the piloting process who were also to be enrolled in the clinical trial in order to have sufficient numbers to conduct reliability testing. The possibility of memory effects with the questionnaires was expected to be minimal as the piloting process occurred 6 months before the questionnaires were used in the clinical trial. The purpose of reliability testing was to test the reliability of respondents in answering the knowledge questions by interviewing children and carers on two occasions 5 weeks apart, Time A and Time B. During the 5 week interval, the dietitian did not provide any additional nutrition and enzyme information to the subjects and their families in an attempt to ensure that knowledge remained constant.

Responses were recoded as correct or incorrect for analysis. Responses to questions of "don't know" were recoded as incorrect in the knowledge questionnaires, and as inappropriate in the self-management questionnaires, because it was considered reasonable to expect respondents to know the answer to the questions. The open-ended question in the children's knowledge questionnaire was coded as either correct, partially correct or incorrect.

Responses to the open questions in the self-management questionnaires were coded as either appropriate or inappropriate. A response was judged to be appropriate if the subject responded to a correct option or did not respond to an incorrect option.

The results for Time A and Time B administrations of the questionnaires were cross-tabulated to assess the magnitude of the change in the response given by respondents for each question. The question pool was improved by deleting or altering questions with greater than 20% change in answers from Time A to Time B, or if greater than 90%, or less than 10%, of respondents gave correct or incorrect answers.

The piloting process also involved submitting the pilot questionnaires to dietitians working at Australian CF centres in order to establish content validity.

Results

Pre-testing

Pre-testing indicated that children had difficulty with questions related to energy. The children appeared not to understand the energy concept and the interviewers reported that one question seemed stilted due to the use of three related terms, namely energy/kilojoules/calories. Some children also had difficulty determining which of two stated foods contained the most salt. Two carers stated that they did not know the answer to the question about foods suitable for mixing enzyme granules into because they had never had to do this. One carer had difficulty realising that two questions were asking different things about the same concept, ie. the first question asked about the age for 'no' supervision and the second question about the age for 'some' supervision. Some carers found that the set of questions which asked about the signs of malabsorption was difficult as the options included signs which could be due to other conditions, such as gastroenteritis.

An interviewer suggested that the question for children of "Should children with CF cut down on how much salt they have?" be changed to "Should children with CF stop eating salty food?" in order to avoid misinterpretation.

The interviewers indicated that the levels of respondent interest and attention were appropriate and that the interviews were not too long for the topics assessed. The interviewers suggested that the flow of the questionnaires could be improved by preceding some sets of questions by an introductory explanation. For example, "Soon I will be asking questions about the energy, kilojoules and calories you get from eating and drinking." Repetition of part of the question was also suggested by the interviewers for the three sets of questions regarding which of two foods has the most fat, energy and salt. For example, "I'm going to read a list of foods. Of each pair which has the most fat: a meat pie or a banana, which has the most fat?"

Pre-testing also indicated the need for several other improvements to the questionnaires. The improvements included: a review of some of the knowledge questions which appeared to the interviewers to be too simple; the addition of 'don't know' as an option to the several sets of questions regarding which of two foods has the most fat, energy or salt, so as to discourage guessing; instructions for interviewers to direct respondents to select only one year of age, not a range of ages, for relevant questions; and typing errors that needed correcting.

Piloting of the other instruments to be used during the clinical trial with respondents involved in pre-testing indicated that the interviewer could personalise the questions in the behaviour questionnaire by establishing the gender of the carer's child/ren with CF at the commencement of this section of the interview. An interviewer also suggested that the word 'negotiate' in one of the behaviour questions be altered to 'discuss' for better comprehension. The household record form, energy expenditure recording sheet and food frequency questionnaires did not appear to require any alterations.

Piloting

Reliability testing revealed that 3 of the 38 questions in the children's pilot knowledge questionnaire, and 18 of the 53 questions in the carers' pilot knowledge questionnaire, needed to be deleted or altered as they were non-discriminatory or there was greater than 20% change in respondents' answers over time, most likely due to guessing.

Based on these results, one question in the children's knowledge questionnaire was deleted. Thirteen of the questions in the carers' knowledge questionnaire were deleted, two new questions were included and five questions were altered. Two questions in the children's questionnaire and one question in the carers' questionnaire were not deleted or altered as the poor response was considered to be due to a lack of necessary knowledge rather than a problem with the questions. For example, the question: 'Does cheese have lots of salt?' was not deleted.

Questions that were deleted included: those testing basic knowledge, such as enzymes are needed with milk and that the number of enzymes needed depends on the amount of fat consumed; those assessing foods that respondents were not familiar with, such as clear soup; and ambiguous questions, such as does coughing use up a lot of energy. Respondents indicated that their answer would depend on the child and amount of coughing. Alterations included changing questions such as "Which has the most fat: a meat pie or a banana?", to "a boiled egg or a banana?"

The interviewers indicated that the questions about energy needed an introduction that was even more detailed in order to explain the kilojoule/calorie content of food concept more clearly.

Piloting indicated that the self-management questionnaire could be improved by changing 'would you' to 'could you' in relevant questions in order to overcome respondents indicating their preferred action versus their knowledge of the options available.

Six out of 13 dietitians who were approached by mail to review the questionnaires returned the instruments along with their comments. Their expert opinions indicated that, with a few minor alterations, the questionnaires were suitable for measuring the knowledge and self-management requirements of children with CF and their carers. An example of a suggestion for enhancing content validity was to alter the question "Can tummy aches occur if enzymes are not taken with food?" to "Could you get a tummy ache if you forgot to take your enzymes with a meal?" It was suggested that respondents may think of fruit as the food in the original question and fruit is one of few foods which does not require enzymes. Other recommendations included: changing the question about taking enzymes 'before a meal' to 'with a meal' to decrease confusion, given that the recommendation is to take enzymes both before and during a meal; deleting the question about the energy value of clear soup as respondents may be unfamiliar with the item; incorporating an assessment of how children would respond to being teased at school when taking enzymes; explaining

what malabsorption is before asking questions about dealing with the problem; changing 'checking weight' to 'checking growth' so as to be promoting a positive body image and not focussing only on weight; dividing the time for improvement in bowel actions from 1 to 7 days into two options; changing questions on how carers might deal with enzyme and food refusal to open-ended questions with a rating scale as respondents may be reluctant to admit to negative actions like punishing a child or getting angry; grouping all questions about the same topic (eg. enzymes) together; and assessing knowledge about the fat content of mixed meals.

Discussion

Pre-testing

Pre-testing the questionnaires before piloting enabled improvements to be made based on feedback obtained from respondents and interviewers. All of the problems that were identified during pre-testing were addressed, most of the suggested changes were made and some questions were deleted. The alterations that were made to the questionnaires included changing the set of questions which asked about malabsorption to signs of 'possible' malabsorption; transition statements were inserted between each domain to help promote interest and cooperation from the respondent (eg. "Soon I will be asking you questions about salt in food. I'm going to read a list of foods. Of each pair which has the most salt...?" and the introduction "Which of the following would help a child with CF increase their energy intake?" was changed to "Soon I'll be asking you questions about energy kilojoules and calories you get from eating and drinking. Which of the following....?"); and brief introductions and explanations of complex terminology, such as malabsorption, were included to enhance understanding. The two changes that were not made were the suggestion to use only one of the terms related to energy intake (energy, kilojoules or calories) to enhance fluidity of the questions and the suggestion to decrease the bias associated with the use of pictures to illustrate some questions for children. Piloting of the questionnaires had indicated that individual respondents may be familiar with different energy terms. Rather than use only one term, respondents were asked to indicate which term they were most familiar with and this term was then used for these types of questions (Appendix 3.2 and 3.3). The suggestion that the picture file

introduced bias was considered to be unjustified as foods illustrated for individual questions were all the same size and colouring. Pre-testing was considered to be a very important process for developing the questionnaires and for enhancing validity. Other instruments which have been used to assess CF nutrition and pancreatic enzyme knowledge and self-management skills do not appear to have undergone pre-testing of piloting (91, 93, 94).

Piloting

Content validity of the nutrition and PERT knowledge and self-management questionnaires for children with CF and their carers was initially established by basing the questions on the learning objectives of the *Go and Grow with CF* nutrition and behaviour intervention program (Section 3.2) and then by submitting the pilot questionnaires to dietitians working at Australian CF centres. The validation process indicated that, with a few minor alterations, the questionnaires were suitable for measuring the knowledge and self-management requirements of children with CF and their carers.

Submitting the questionnaires to dietitians with CF expertise was important to determine if the questionnaires asked relevant questions about nutrition and PERT and covered sufficient issues. A question about taking enzymes in front of other children was included in the children's self-management questionnaire as suggested by a dietitian who reviewed the pilot questionnaires. Such a question is very relevant to an aim of the intervention, that being to increase children's confidence with treatment.

Reliability testing during piloting enabled questions in the knowledge questionnaires that were poor discriminators and unlikely to detect a difference between groups to be identified. These questions were deleted or altered. Suggestions from interviewers and dietitians were also used to improve the questionnaires (eg. all questions of a similar domain, such as fat, were grouped to aid concentration).

Reliability testing of the same questionnaires indicated that not all of the dietitians' concerns were valid. For example, carers were willing to admit to negative actions like getting angry when their child refused food. Therefore, these response options were not deleted and the question was left as closed-ended. The suggestion by a dietitian to assess knowledge about the fat content of mixed meals was not incorporated in the questionnaires as such questions would require extensive pre-testing and reliability testing. A detailed introduction to the energy concept was included by using cars as an analogy of energy needs, ie. cars need petrol to go which is similar to the need of people who need food to give them energy to go.

Two of the children's, and one of the carers', knowledge questions which scored poorly during this reliability testing were retained. The investigators agreed that these questions were phrased well and the poor result was most likely due to lack of knowledge, rather than ambiguity. An example of such a question is, "Does cheese contain lots of salt?"

The final children's knowledge and self-management questionnaires contained 37 and 15 questions, respectively, and took a total of 12 minutes, on average, to administer. The highest possible score for the children's knowledge questionnaire was 37. The highest possible scores for total appropriate and total inappropriate responses to the five scenario-based questions in the children's self-management questionnaire were 23 and 11, respectively (Appendix 3.2).

The final carers' knowledge and self-management questionnaires contained 42 and 17 questions, respectively, and took a total of 20 minutes, on average, to administer. The highest possible score for the carers' knowledge questionnaire was 42. The highest possible scores for the appropriate and inappropriate responses to the twelve scenario-based questions in the carers' self-management questionnaire were 61 and 41, respectively (Appendix 3.3).

3.6 Readability

Methods

Readability of the home-based course, the final knowledge and self-management questionnaires for children and carers and standard written nutrition information (Healthy Eating the CF Way) (128), that is given to parents when their child is diagnosed with CF at Princess Margaret Hospital, was assessed by applying the SMOG (simple measure of gobbledegook) Formula (130). Although clinical and research psychologists with expertise in child development were involved in formulating the intervention program and questionnaires, it was considered useful to assess readability, particularly of the material for children, in order to ensure that the levels were lower than that of the standard written information that is usually given to parents.

The SMOG formula is based on the theory that texts with many polysyllabic words require higher reading comprehension levels than texts with words that are mainly one or two syllables. The score is determined by calculating the average number of polysyllabic words for every 30 sentences. The reading grade that is estimated by the SMOG formula relates to average reading levels among the USA population.

Results

The SMOG scores were as follows:

- 7.5 for the home-based course
- 7.0 for the children's knowledge and self-management questionnaires
- 8.5 for the carers' knowledge and self-management questionnaires, and
- 11 for the standard written nutrition information (128) that is given to parents when their child is diagnosed with CF.

Many of the words in the home-based course and questionnaires that were polysyllabic were names of foods and concepts particular to CF, namely banana, lemonade, potato, energy, malabsorption and infection, and words that directed

participants during the weekly activities of the home-based course, namely remember, following and answering.

Discussion

Consequently, the readability of the home-based course and questionnaires for carers was considered to be appropriate, in that the scores were 7.5 and 8.5, respectively. Hawe et al (130) suggest that health information which is assessed as having a SMOG score of less than 12 should reach a wide Australian (adult) audience. By comparison, the SMOG score of 11 for the standard nutrition information (128) was 3.5 units higher than the home-based course material, indicating that more carers are likely to comprehend *Go and Grow with CF*. McLaughlin (131) recommends aiming for a reading level of 6 or less in order for the majority of adult readers to comprehend health education material. Such a low score was difficult to achieve in the *Go and Grow with CF* program as many of the polysyllabic words were names of foods commonly eaten and concepts particular to CF. Nonetheless, appropriate levels of readability were planned for during the development of *Go and Grow with CF*, an aspect which is often not reported.

The score of 7.0 for the children's knowledge questionnaire justifies the use of an interview for administering the questions and for providing an explanation of CF- and food-related polysyllabic words, such as malabsorption and banana. The score of 7.5 for the home-based course supports the need for carer involvement in completing the material. Children as young as 6 years of age are encouraged to be actively involved in the program even though it is recognised that most would be unable to read and comprehend the information on their own. The design of *Go and Grow with CF* relies on carers to both read the home-based course material with their child, as well as reinforce steps taken toward behaviour change. It was not possible to decrease the number of the polysyllabic words in the home-based course as most were specific to CF or to food commonly consumed and it would have been inappropriate to exclude them.

4. CHAPTER FOUR: THE CLINICAL TRIAL - METHODS

The purpose of the clinical trial was to evaluate the pilot *Go and Grow with CF* program. The methodology and results from the process evaluation of the pilot program and the assessment of its impact on knowledge, self-management, behaviour, dietary intake and body composition, using anthropometry, are presented in this and the next chapter.

4.1 Study design

The clinical trial consisted of baseline, pre- and post-intervention measurements, together with data from 6 and 12 month follow-up. Baseline measurements were taken at least 6 months before the intervention and the time period between pre- and post-intervention measurements was 4 to 6 months. Children were randomly assigned to either the control or intervention group using a random number table.

The intervention being assessed by the clinical trial was participation in the *Go and Grow with CF* program. At the beginnning of the intervention stage, both the intervention and control group were provided with updated, standard written information (128) to control for the possibility of benefits from clinical attendance associated with the *Go and Grow with CF* intervention program. Throughout the clinical trial, both the intervention and control group continued to receive routine one-to-one dietetic counselling.

4.2 Participants

Children eligible for the clinical trial were those attending the Princess Margaret Hospital CF clinic who were aged between 2 and 11 years at the commencement of baseline data collection and who had pancreatic insufficiency. Exclusion criteria for the clinical trial were pancreatic sufficiency, liver disease requiring drug therapy,

short-gut syndrome, enteral-tube feeding and marked language delay in children aged 6 to 11 years. Children with pancreatic sufficiency were excluded as half of the home-based course material covers topics that are not relevant to these individuals, such as enzymes and malabsorption. Liver disease requiring drug therapy, short-gut syndrome and enteral-tube feeding were considered to be exclusion criteria due to their significant effects on nutritional status which would have confounded the results. Children aged 6 to 11 with marked language delay, due to conditions such as autism and cerebral palsy, were excluded as their reduced ability to complete the program and the questionnaires may have confounded the results. Children aged 0 to 2 years were not included in the clinical trial because their carers had participated in a group intervention program in the preceding year. Children aged 12 years and older were excluded because of their different learning needs associated with both the cognitive changes of adolescence (which require different teaching styles to those for younger children) and the increasing influence of peers rather than carers in this age group (although the increasing influence of peers during adolescence is not a reason not to develop programs that focus on both adolescents and their parents).

4.3 Process evaluation

Participation rates were calculated as a proportion of those who agreed to take part in the program. Carers' program goals for themselves, and for their children, were documented during the introductory workshop or after implementation advice was provided by telephone. At the completion of *Go and Grow with CF*, carers indicated if these program goals were achieved. During telephone contact with the dietitian (me) at weeks 4, 7 and 10 of the home-based course, carers self-reported what they, and what they thought their child, were learning, enjoying and disliking by answering open-ended questions (Appendix 4.1). Carers were also asked to indicate the effect of the program on their family; what helped them to continue participating in the home-based course; and whether they would recommend *Go and Grow with CF* to other families who have a child with CF. Many of the process evaluation questions were open-ended. Consequently, percentages are not reported for response categories, as assigning percentages would suggest that carers were given an

opportunity to comment on each category in turn (130). The amount of dietetic time spent conducting all aspects of the program over three months was recorded.

4.4 Knowledge and self-management

Materials

Development and validation of the separate but similar nutrition and PERT knowledge and self-management questionnaires for children and carers is described in Section 3.5. The final questionnaires used in the clinical trial are located in Appendix 3.2 and 3.3. The questionnaires were administered as face-to-face interviews with children aged 6 to 11 years and by telephone with carers of 2 to 11 year olds on three occasions, namely pre-intervention, post-intervention and at the 12 month follow-up time point. The interviewers were dietitians who did not work in the area of CF and were blinded to group allocation in order to minimise the effects of social desirability of responses by respondents and investigator bias. Respondents may have been inclined to guess answers to questions rather than admit that they did not know the answer if the aspects being assessed had been taught by the interviewer.

Statistical analysis

The Statistical Package for Social Sciences 6.1.3, 1995 (SPSS Incorporated, Chicago, Illinois, USA), was used to determine descriptive statistics for the questionnaire responses, to assess the relationships between carers' and children's knowledge, self-management and socioeconomic index scores and to assess the effects of the intervention. Comparisons were performed using independent t-tests.

Total percent knowledge, and appropriate and inappropriate self-management scores were determined for the children and carers' questionnaires. This was done by summing the number of correct answers for the knowledge questionnaires and the number of appropriate and inappropriate responses for the self-management questionnaires. A score was also calculated for each domain in the knowledge questionnaires. The frequency of unprompted responses alone, and of the

unprompted and prompted responses combined, was determined for the self-management questions.

The average differences in the scores from pre- to post-intervention and from pre-intervention to the 12 month follow-up time point were calculated for participants' within each group to assess both the immediate and long-term changes. Independent t-tests were then performed on participants' differences in scores over time to compare the difference in the average improvement between the intervention and control groups.

In addition to calculating an overall score for the children's and carers' self-management questionnaires, each question was also analysed individually. Changes over time were assessed and the differences in the change between groups were analysed by:

- evaluating the proportion of unprompted alone, and combined unprompted and prompted, responses that were appropriate for the intervention group and determining whether the proportion had improved or declined over time
- determining, by cross-tabulation, the proportion of participants who gave only appropriate responses, and
- performing independent t-tests on the individual differences over time on the total of appropriate and inappropriate responses for each question.

Associations between the knowledge scores of the children and carers, and the changes in the scores, in each group over time, were examined using Pearson's correlation coefficient. Spearman's rank correlation was used to examine the associations between children's and carers' knowledge and self-management scores and socioeconomic index as the scales of measurement in the questionnaires differed (eg children's maximum possible score for the knowledge questionnaire was 37 and for the self-management questionnaire was 23 appropriate responses and 11 inappropriate responses).

The effects of socioeconomic index on group and mean knowledge and self-management scores were assessed using chi-square analysis and one-way analysis of variance.

4.5 Behaviour

Materials

Following on from the pre- and post-intervention and 12 month follow-up knowledge and self-management telephone interview, all carers were also interviewed using a modified form of the pre-formulated and validated Behavioural Pediatrics Feeding Assessment Scale (BPFAS) (Appendix 4.2).

The BPFAS was developed by Crist et al (106) to assess the meal-time behaviour of children with CF and their carers. Modification of the BPFAS for this clinical trial involved changing the questions from present to past tense. The questionnaire contained 35 items which focussed specifically on refusal to eat. The first 25 items were descriptions of the child's behaviour, and the next ten questions described parental feelings or strategies (responses) for dealing with eating problems. Each item presented a descriptive phrase for which the carer rated how often the behaviour occurred on a 5-point Likert scale from "never" to "always" and whether or not the carer considered the behaviour to be a problem ("yes" or "no"). Questions were phrased both positively and negatively (eg. my child will try new foods and my child cries at feeding time). The interviewers were dietitians who did not work in the area of CF and were blinded to group allocation in order to minimise the effects of social desirability of responses by respondents and investigator bias. Respondents may have been inclined to deny the presence of food-related behaviour problems if the interviewer was a health-care provider to whom they had denied such problems.

Statistical analysis

The behavioural data examined were the scores generated for both sections of the questionnaire, child and carer's, from both the Likert scale responses (frequency ratings) and from the number of behaviours endorsed as problems. When the

protocols were scored, ratings were transformed so that higher scores meant more behavioural problems.

The Statistical Package for Social Sciences 10.0, 1999 (SPSS Incorporated, Chicago, Illinois, USA) was used to determine descriptive statistics for the scores, to assess differences between the groups and between genders immediately prior to the intervention and to assess the effects of the intervention. All comparisons were made using independent t-tests. Longitudinal effects on frequency scores were assessed by calculating the average difference in the scores for subjects within each group from pre- to post-intervention and from pre-intervention to 12 month follow-up (to assess both the immediate and long-term changes). Independent t-tests were then performed on subjects' differences in scores over time to compare the difference in the average improvement between the intervention and control groups.

The SAS System 8.0, 1999 (SAS Institute Inc, Cary, North Carolina, USA) was used to assess the longitudinal effects of the intervention on problem scores using a logistic regression model. The child and carer's problem scores were declared as binary outcome variables (ie, no problems or 1 or more problems) since many respondents indicated that they had no problems with the child behaviours and carers' responses. Time and group were declared as class (categorical) variables, with three and two categories. Correlation between binary responses on the same individual over the three time points was accounted for using a generalised estimating equation approach with an exchangeable correlation structure, which assumes that the correlation between all outcomes on a single individual are equally correlated.

4.6 Dietary intake

Materials

The meal-based quantitative FFQ, developed and validated for adults by the Deakin Institute of Human Nutrition as part of the Geelong public health nutrition study

(GPHNS) (61, 64), was used to assess the dietary intake of children with CF at each of the five time points. The open-ended GPHNS FFQ contains a comprehensive prompt list of over 300 foods and requires a response only for foods actually eaten and for the relevant meal at which it was eaten (ie. breakfast, lunch, dinner and/or between meal snacks). Items in the questionnaire are arranged by food group and the serve size is described in household measures. The frequency of consumption of a specified amount of each item is requested either as per month, week or day, rarely (when a food was consumed less than once per month) or as never eaten. An open-ended question was asked at the end of the questionnaire to probe for foods that may have been eaten but which were not listed. Additional questions regarding methods of food preparation, types of foods eaten and additions of fat and salt were also included (Appendix 4.3). The additional information was used during analysis to supplement and quantify the dietary data listed in the response booklet.

FFQs were either posted or given at CF clinic appointments to carers, to complete on behalf of the 58 children enrolled in the clinical trial, during each of the five data collection periods. Although children aged 10 years and over may be able to give accurate dietary information (132), carers completed FFQs on behalf of all the subjects in this study in order for the characteristics of the respondents (adults vs children) to be consistent. Also, dietary data obtained directly from children may be limited by children's variability in comprehension and concentration skills (63), poor memory of dietary intake and insufficient knowledge of food and food preparation (58).

At baseline, carers received verbal instructions for completing the FFQ either at the CF clinic or by telephone. Written instructions for completing the FFQ were also provided during each of the five data collection periods (Appendix 4.3). These instructions were developed by Wright and Scott (133), based on the questionnaire sample page, and emphasised that this was an assessment of the child's intake over the previous 3 months. Carers returned completed FFQs by post in a pre-paid return envelope. The questionnaires were checked for completeness and any queries investigated with the carer by telephone. After each of the five data collection

periods it was necessary to contact a few carers who had not returned the FFQ booklets within 1 month.

Dietary analysis

Nutrient analysis was carried out using SOFFA (System for On-line Food Frequency Analysis, version 1.0b. Cottesloe, Western Australia: Computer Models; 1992) and based on the Australian nutrient composition database NUTTAB91 (Nutrient data tables for use in Australia. Canberra: Commonwealth Department of Community Services and Health; 1991). Two dietitians, who were blinded to subjects' group allocation, coded all the FFQ responses using a pre-determined coding protocol. Consistency of coding and data entry were checked regularly on subsets of the FFQ.

Although it has been recommended that the plausibility of energy intakes from dietary assessment methods should be assessed in order to identify under-reporters, by comparing energy intake to basal metabolic rate (134), the investigators of this study considered it inappropriate to exclude any of the subjects from analysis. Firstly, the cut-off points for under-reporting may not be appropriate to use when examining the data of children as the Goldberg data (134) are from studies conducted with adults. Also, it would be impossible to determine a correction factor for under- or over-estimation of actual intake unless CF-related energy expenditure and losses of each subject were measured or comparisons made with a superior method of dietary recording.

Statistical analysis

The Statistical Package for Social Sciences 10.0, 1999 (SPSS Incorporated, Chicago, Illinois, USA) was used to assess the dietary intake measurements of children with CF. The data examined included energy (MJ), protein and fat (g), energy as %RDI, protein and fat as a percentage of total energy intake, sodium (g) and zinc (mg). Sodium intake was assessed, in addition to the nutrients known to affect growth, as individuals with CF have elevated requirements due to excessive sweat losses (Section 2.6). Analysis involved determining descriptive statistics for the nutrients, comparisons between groups and genders immediately prior to the intervention using

independent t-tests and assessments of the effects of the intervention using multivariate repeated measures analysis.

Although the subjects had been randomly allocated to the intervention and control groups, the data were also analysed using gender, genotype, PERT, socioeconomic index, illness and activity levels as additional independent factors, and measures of lung function as covariates, to statistically control for any possible effects (Sections 4.8 to 4.13).

4.7 Body composition

Measurements

Subjects were measured at all five time points by anthropometrists and respiratory technicians blinded to group allocation, using standardised techniques and procedures (135). Measurements were recorded using a form that was in accordance to these standards (Appendix 4.4).

All subjects were weighed in their underclothes on a digital scale (Wedderburn, NSW, Australia) to the nearest 0.1 kg. Stretched stature was measured in triplicate with a stadiometer (Holitan, Crymych, England) to the nearest 0.1 cm and the median value recorded. Self-reported heights of the biological parents were obtained from the subjects' mothers.

Initially, weight and height z-scores were determined for the 2 to 11 year olds using the Ozgrow Software Program, 1995 (Camperdown, Australia) which compared the subjects' measurements to the National Center for Health Statistics (NCHS) reference group of North American children (136). Percent ideal body weight (%IBW) was calculated based on the subjects' observed NCHS height-for-age percentile (18). The genetic contribution to linear growth was evaluated using the parental height-adjustment method of Himes et al (137).

Additional anthropometric measurements were taken of the children aged 5 to 11 years. Anatomical landmarks were identified on each subject and skinfolds and girths were measured by one of two anthropometrists, according to the methods prescribed by Ross & Marfell-Jones (135). Calipers (Slimguide, Plymouth, England) were used to measure 8 skinfolds (triceps, subscapular, biceps, iliac crest, supraspinale, abdominal, front thigh and medial calf) to the nearest 0.5 mm. A flexible tape (Lufkin, USA) was used to measure 10 girths (upper arm relaxed, upper arm flexed and tense, forearm, wrist, chest, waist, gluteal, thigh, calf and ankle) to the nearest 0.1 cm. Both skinfolds and girths were measured in triplicate and the median value recorded.

The two anthropometrists also measured six children without CF and technical error of measurement scores were calculated for each skinfold and girth. The technical error of measurement score was determined in order to assess each anthropometrist's level of precision. The technical error of measurement score is equivalent to the coefficient of variation of the measurement and is calculated as the within subject ratio of the standard deviation to the mean and expressed as a percentage (138). Based on the data obtained from the six children without CF, the mean technical error of measurement scores of the skinfolds measured by anthropometrists 1 and 2 were 6.4% and 7.0%, respectively (target range $\leq$ 5%) and between anthropometrists was 8.4% (target range $\leq$ 7.5%). The mean technical error of measurement scores of the girths measured by anthropometrists 1 and 2 were 0.8% and 0.9%, respectively (target range $\leq$ 1%) and between anthropometrists was 1.8% (target range $\leq$ 1.5%) (139).

The primary anthropometric measures determined from the skinfold and girth measurements, and used in both the cross-sectional and longitudinal analysis, were muscle mass (MM) (derived from 4 skinfolds, namely triceps, subscapular, front thigh and medial calf, and 5 girths, namely arm, forearm, chest, thigh and calf) (140), mid-upper arm circumference (MUAC) and arm muscle area (AMA) (derived from the triceps skinfold and upper-arm girth) (141), adipose mass (AM) (derived from 6 skinfolds, namely triceps, subscapular, supraspinale, abdominal, front thigh and

medial calf) (140), percent body fat (%fat) (derived from triceps and either calf or subscapular skinfolds) (142) and the sum of the 6 skinfolds (Σ6SF) (derived from triceps, subscapular, supraspinale, abdominal, front thigh and medial calf skinfolds). The longitudinal analysis of the data also included secondary anthropometric measures, namely the sum of 4 skinfolds (derived from triceps, subscapular, biceps and supraspinale skinfolds) and the sum of 8 skinfolds (derived from triceps, subscapular, biceps, iliac crest, supraspinale, abdominal, front thigh and medial calf skinfolds).

A second set of age- and gender-standardised z-scores was determined for 5 to 11 year olds by comparing the subjects' height, weight, skinfold and girth measurements to those of children of the same age in the Coquitlam growth study (COGRO) reference group (143).

The North American NCHS and Canadian COGRO reference groups were used in this study because comprehensive Australian data are not available. The NCHS reference group was used for initial height and weight comparisons since growth charts based on the data are frequently used in the clinical setting. The NCHS data are measurements made on large, nationally representative samples of children living in the USA between 1963 and 1975 (144). The anthropometric data of the COGRO reference group was also used for comparison since the NCHS data does not include skinfold and girth measurements. The COGRO reference group included middle class children from the west coast of Canada who were aged 5 years or more (n=316 were 5 to 11 years of age) (143). It should be noted that the COGRO reference group is likely to have been fitter than the NCHS reference group, as the schools from which the COGRO children were drawn encouraged a high level of physical activity.

Our cross-sectional analysis of the skinfold and girth measurements was based on data from 31 children aged 6 to 11 years only, so that the results would be suitable for comparisons with future studies which are not likely to include children less than 6 years of age as they are mostly uncooperative with the measurements.

Longitudinal analysis of the skinfold and girth measurements includes the children who were 5 years of age at baseline.

Statistical analysis

The Statistical Package for Social Sciences 6.1.3, 1995 and 10.0, 1999 (SPSS Incorporated, Chicago, Illinois, USA) was used to assess the anthropometric and lung function measurements of children with CF. The data examined included height, weight, skinfolds, girths and anthropometric measures (derived from skinfolds and girths). Analysis involved determining descriptive statistics for the measurements, comparisons between genders and between the children with CF and reference groups immediately prior to the intervention, using independent t-tests, and an assessment of the effects of the intervention using multivariate repeated measures analysis in order to account for the five assessment time points of the two groups.

Although the subjects had been randomly allocated to the intervention and control groups, the data were also analysed using gender, genotype, PERT, socioeconomic index, illness and activity levels as additional independent factors, and measures of lung function as covariates, to control statistically for any possible effects (Sections 4.8 to 4.13).

4.8 Genotype

The CF gene mutations of the children with CF were classified as either severe, mild or variable according to the pancreatic dysfunction categories suggested by Durie et al (145).

4.9 Pancreatic enzyme replacement therapy

The children with CF took their usual pancreatic enzyme preparation (either Pancrease of Cotazym-S-Forte) throughout the clinical trial.

4.10 Socioeconomic status

Information regarding socioeconomic status was collected using a household record form (Appendix 4.5), which was based on the Western Australian Child Health Survey Household Record Form and Family Background Questionnaire (146). Subjects were assigned a socioeconomic index which was one of five levels of socioeconomic status, based on their carers' level of education, the size of the family and total family income (146).

4.11 Illness

An average illness level (low, moderate, high) for each subject was determined using data regarding the use of steroids and antibiotics over the five data collection periods (Appendix 4.6).

4.12 Activity

An average activity level (low, moderate, high) for each subject was determined using data regarding the frequency and duration of exercise and chest treatments over the five data collection periods (Appendix 4.7). The subjects' use of oral, inhaled and IV antibiotics and steroids during the clinical trial were recorded. An average illness level was determined based on the frequency and type of medications taken.

4.13 Lung function

Standard spirometry (147) was performed by children aged 6 to 11 years at all time points during the study. Forced expiratory volume in 1 second (FEV_1), forced vital capacity (FVC) and forced expiratory flow at 25 to 75 % of vital capacity

($FEF_{25\% \text{ to } 75\%}$) were compared with age-, gender- and height-specific reference values and reported as a percentage of the predicted value (148, 149) (Appendix 4.6).

Principle component analysis was used to determine linear combinations (ie effectively weighted averages) for %predicted FEV_1 and $FEF_{25\% \text{ to } 75\%}$ measurements for use in the multivariate analyses (Sections 4.6 & 4.7). As the values were highly correlated, the first three principle components of FEV_1 and $FEF_{25\% \text{ to } 75\%}$ were included as covariates in the linear statistical model as they explained over 90% of the variation in the corresponding series of lung function measurements. The principle components of FEV_1 were included in the linear statistical model separately from the principle components of $FEF_{25\% \text{ to } 75\%}$.

Informed written consent was obtained from the carer most responsible for the child's nutrition and pancreatic enzyme therapy. Approval for the intervention program and clinical trial was obtained from the Ethics Committees of Princess Margaret Hospital and Curtin University of Technology.

5. CHAPTER FIVE: THE CLINICAL TRIAL - RESULTS & DISCUSSION

5.1 Participants

Of the 88 children aged 2 to 11 years attending the Princess Margaret Hospital CF clinic, 70 were eligible for the clinical trial. Of these, 11 were not enrolled in the study as they did not attend the clinic regularly or their parents did not consent to their participation. Thus, 59 children aged 2 to 11 years and their primary carer were enrolled in the clinical trial designed to assess the impact of the *Go and Grow with CF* program.

Of the 59 children aged 2 to 11 years who were enrolled in the clinical trial, 30 children and their carers were randomly assigned (using a random number table) to participate in the intervention program. However, one carer and child chose not to participate in the intervention program due to time constraints and their data were excluded from the analysis. Consequently, program participants included 21 children aged 6 to 11 years and 27 carers of 29 children aged 2 to 11 years. Another carer and child, who had been randomly allocated to the control group, transferred interstate and were unavailable for follow-up at 6 and 12 months. Thus, data available for analysis included:

- 58 children aged 2 to 11 years (42 children were aged 6 to 11 years) and 55 carers of 2 to 11 year olds at the baseline, pre-intervention and post-intervention time points, and
- 57 children aged 2 to 11 years (41 children were aged 6 to 11 years) and 54 carers at 6 and 12 month follow-up.

The number of children at the beginning of the clinical trial represented 66% of all children in the 2 to 11 year old age range attending the Princess Margaret Hospital CF clinic. All the subjects were pre-pubertal prior to the intervention, as determined by Tanner staging which was conducted by the CF clinic medical staff (150).

Tables 5.1a & 5.1b show that allocation of group using the random number table method produced two groups with similar characteristics prior to the intervention in terms of age, pulmonary function, height and weight, gender, genotype, PERT, socioeconomic status, illness and activity. Further analysis of the measures of lung function revealed that more females than males had lower than normal levels of %predicted $FEF_{25\% \text{ to } 75\%}$ and FEV_1, but these differences did not reach statistical significance.

Table 5.1a

Characteristics of the children with cystic fibrosis immediately prior to the intervention

		Intervention group n=29[g] mean±sd (range)	Control group n=29[h] mean±sd (range)	Estimate (95% CI)[i]	P-value[j]
Age	Years	7.1±2.6 (3.5 to 11.8)	6.9±2.9 (2.4 to 11.5)	0.2 (-1.3, 1.6)	0.83
Age CF diagnosed	Months	9.8±16.3 (0 to 60)	4.8±5.9 (0 to 30)	5.0 (-1.4, 11.5)	0.12
Lung function[a]	FEV_1[d]	86±19 (53 to 125)	82±14 (56 to 111)	4 (-7, 16)	0.44
	$FEF_{25\ to\ 75\%}$[e]	62±26 (24 to 110)	59±27 (30 to 144)	3 (-16, 22)	0.75
	FVC[f]	99±17 (60 to 120)	93±15 (69 to 133)	6 (-5, 17)	0.30
Weight[b]	z-score	-0.6±0.9 (-2.5 to 1.3)	-0.6±0.9 (-2.8 to 0.8)	0.0 (-0.4, 0.6)	0.70
Height[b]	z-score	-0.4±1.0 (-2.9 to 1.3)	-0.7±1.0 (-3.6 to 1.5)	0.3 (-0.3, 0.8)	0.34
IBW[c]	%	98±9 (79 to 119)	101±9 (82 to 127)	-3 (-8.2, 1.7)	0.19

[a] percent predicted lung function measurements of 6 to 11 year olds, standardised for age, gender and height (148, 149)
[b] standardised for age and gender (136)
[c] percent ideal body weight (18)
[d] forced expiratory volume in 1 second, normal range >85%
[e] forced expiratory flow at 25% to 75 % of vital capacity, normal range >65%
[f] forced vital capacity, normal range >85%
[g] n=16 for lung function, as measurements taken only of 6 to 11 year olds
[h] n=17 for lung function, as measurements taken only of 6 to 11 year olds
[i] estimate (confidence interval) for the difference between groups
[j] independent t-test for the difference between groups

Table 5.1b

Characteristics of the children with cystic fibrosis

		Intervention group n=29[f] n (%)	Control group n=29 n (%)	χ^2 (d.f.)[g]	P-value
Gender	Males	17 (59)	14 (48)	0.62	0.43
	Females	12 (41)	15 (52)	(1)	
Genotype[a]	1	23 (85)	21 (74)	1.36	0.24
	2	4 (15)	8 (26)	(1)	
	3	0	0		
PERT[b]	Pancrease	19 (66)	15 (52)	1.14	0.29
	Cotazym-S-Forte	10 (34)	14 (48)	(1)	
SEI level[c]	1	3 (10)	4 (14)		0.69
	2	4 (14)	5 (17)	2.27	
	3	12 (42)	9 (31)	(4)[h]	
	4	9 (31)	11 (38)		
	5	1 (3)	0		
Illness[d]	1	1 (3)	5 (17)	3.24	0.20
	2	17 (59)	15 (52)	(2)[h]	
	3	11 (38)	9 (31)		
Activity[e]	1	4 (14)	7 (24)	1.19	0.55
	2	14 (48)	11 (38)	(2)[h]	
	3	11 (38)	11 (38)		

[a] 3 CF gene mutation categories (severe, mild and variable) (145), analysis computed only for the resulting 2 x 2 table
[b] pancreatic enzyme replacement therapy
[c] socio-economic status index, 5 categories (high to low) based on qualifications and education level of both carers of the child and total family income
[d] 3 levels (low to high) based on the average usage of antibiotics and steroids over the 5 time points
[e] 3 levels (low to high) based on the average level (frequency and duration) of exercise and chest treatments over the 5 time points
[f] n=27 for Genotype
[g] Pearson chi-square test statistic for contingency tables
[h] Likelihood ratio test statistic used, due to some cells having an expected count <5 (degrees of freedom)

5.2 Process evaluation

Methods (Section 4.3)

Results

Participation: Only 57% of children and 50% of carers attended both workshops. Ten percent of children and 30% of carers did not attend either workshop. Carers' reasons for not attending the workshops were related to not wanting to attend extra hospital activities, having work commitments or family illness. Seventy-three percent of children and/or their carers completed the home-based course on schedule, ie before the concluding workshop. Family illness, such as chickenpox and being busy, temporarily interrupted 10% of families. Three weeks after the concluding workshop a total of 82% of children and/or their carers had completed the home-based course. Results of the process evaluation are reported for the group of 82% of children and/or their carers who completed the home-based course. Reasons given by carers who did not complete the home-based course were related to being too busy or having extra demands on the carer's time from newly-born or disabled siblings. All these carers expressed the desire to participate in the program at a later stage.

Goals: The goals carers set for themselves and for their children were related to nutrition and/or pancreatic enzyme requirements of a child with CF. The following is a summary of the most common goals that carers set.

For themselves:

- to learn more about food, nutrition, enzymes, malabsorption
- to help their child gain weight, eat faster, keep healthy, avoid bowel blockages
- to manage their child's diet better, be more adaptable with their child's food, be one step ahead of their child in the area of food
- to learn how to deal with nutrition and enzyme problems, and
- to learn how to maintain their child's interest in treatment, to help their child understand CF-related health needs and be satisfied that their child has been informed about CF needs.

For their children:

- to learn more about CF nutrition needs, high fat foods, enzymes and their importance and malabsorption
- to be more responsible about nutrition and enzyme needs, and
- to remember to take enzymes.

Ninety-six percent of carers reported achieving the goal they set for themselves. Eighty-five percent of carers reported that the goal they set for their child was achieved.

Learning: Ninety-one percent of carers reported learning at least one aspect from a range of topics, including pancreatic enzymes, foods high in fat and salt, increasing energy intake and malabsorption. Ninety-three percent of carers perceived that their children were learning from the program. Following is a list of the nutrition and enzyme concepts that carers reported they and their children learned.

Concepts learned by carers

- the correct use of enzymes and that enzymes are most active for 30 minutes
- what foods are high in fat and how to increase energy intake
- the need for salt and what foods are high in salt
- the signs of malabsorption, and
- reinforcement about all topics and requirements placed into perspective.

Concepts learned by children

- the correct use of enzymes and the importance of taking enzymes
- the signs of malabsorption
- the need for food high in fat, and
- the need for salt and the foods that are high in salt.

Enjoyment: Ninety-one percent of carers enjoyed at least one aspect about the course, such as helping their child learn, learning themselves or increased confidence from a better understanding of nutrition and PERT. Aspects of the home-based course that carers reported children most enjoyed were the rewards for participation

each week, monitoring every day (ticking boxes, drawing smiley faces) and spending time with their carer. Rewards which were most successful in maintaining participation in *Go and Grow with CF* varied. Responses were equally spread over activities (sleep overs, board games, computer time), objects (stickers, toys), money, and food treats (fast food, lollies). Some children were not interested in material rewards. These children enjoyed daily monitoring of their progress on the worksheets or spending time with their carer doing the program.

Dislikes: During the home-based course, carers reported they disliked being too busy to easily fit the program into the week, daily recording and not having planned food available. Thirty-six percent of carers could not identify aspects they disliked about the program. Some carers thought children disliked their play-time being interrupted to do the worksheets and daily recording.

Effect on the family: Fifty percent of carers reported that *Go and Grow with CF* had a positive effect on the family, such as sibling interest in CF nutrition and PERT needs. Nine percent of carers reported that the program had a negative effect on the family, such as a sibling requesting high energy, high salt food at the beginning of the home-based course. Carers who indicated that *Go and Grow with CF* had no effect on the family did not offer any explanation as to why.

Motivation: Telephone calls, which lasted 20 minutes, on average, and fortnightly newsletters from the dietitian motivated carers (95% and 62%, respectively) to continue participating in the home-based course.

Recommendation: All carers who participated in the program indicated they would recommend *Go and Grow with CF* to other families with a child who had CF. Following is a summary of carers' reasons for recommending the program, which include benefits related to both knowledge gain and behavioural change that occurred in them and/or their children.

Knowledge gained

- much was learned from the program that was not known before
- the program was helpful with young children and especially soon after diagnosis
- children learnt about nutrition and enzyme needs
- doing the program was a good process for revision
- the program was more informative than previous information
- the program was set out in plain language, and
- increased awareness of what children are eating and what they need added.

Behaviour changed

- improved habits and involved the child
- helped carers make the effort to provide appropriate food, and
- helped carers work out nutrition and enzyme problems.

Dietetic time: An average of two and half hours per family was spent by the dietitian conducting the **Go and Grow with CF** program over three months. The time was divided between workshops, telephone contact, newsletters and providing feedback on completed worksheets.

Discussion

The process evaluation of **Go and Grow with CF** indicated a high rate of participant satisfaction and perceived learning. Workshops provided a group learning experience and enabled the dietitian to orientate carers to concepts in the home-based course. The low workshop participation rates did not appear to limit the completion rate of the home-based course. Half of the carers who did not attend either workshop did complete the home-based course. Data obtained from the process evaluation questionnaire (Appendix 4.1) suggested that factors other than the workshops provided sufficient motivation for carers to fit the program into their busy schedules each week. These motivational factors are likely to have been the desire of the carers to learn, the flexibility of the program, monthly telephone contact with the dietitian and receiving fortnightly newsletters. A few carers appeared to enjoy sharing highlights during the telephone calls regarding what they and/or their child had

learned. Some carers volunteered that the expectation that their progress with the program would be assessed during the telephone calls was a helpful motivational factor.

Involvement in the program enabled almost all the carers' goals for themselves and their children to be met. Carers recognised that some of the goals that were not achieved were inappropriate for the young age of the child (eg. wanting a five year old to always remember to take enzymes).

The results obtained from process evaluation using the questionnaire located in Appendix 4.1 indicated that all carers would recommend the *Go and Grow with CF* program to other families with a child who has CF. The results also indicated that this high level of recommendation is most likely due to the majority of carers having perceived that they, and their children, learned new concepts from *Go and Grow with CF* and that carers enjoyed the learning process.

5.3 Knowledge and self-management

Methods (Section 4.4)

Results

Pre- and post-intervention data available for analysis included all 42 children with CF aged 6 to 11 years and 55 carers of 2 to 11 year olds. At 12 month follow-up the data available was of 41 children and 54 carers.

Cross-sectional

Knowledge Questionnaire - Children

Pre-intervention, the children's mean knowledge score was 63%. Questions about enzymes and fat were answered better than those regarding salt and energy (Table 5.2). Performance on individual questions in the knowledge questionnaire is shown in Table 5.2.

Table 5.2

Frequency of correct responses to nutrition and enzyme knowledge questions obtained by children with cystic fibrosis aged 6 to 11 years (n=42) and carers of 2 to 11 year olds (n=55)

Questions	Children % with correct response	Carers % with correct response
Enzymes		
What do enzymes do? (digest food) [a]	33	n/a
Arc enzymes needed when eating a biscuit? (Y)	86	94
Are enzymes needed when drinking milk? (Y)	74	n/a
Are enzymes needed when eating an apple? (N)	77	94
Are enzymes needed when eating cake? (Y)	95	100
Do enzymes work best if taken after eating? (N)	79	86
When you eat a fatty meal should you take more enzymes? (Y)	77	n/a
Could you get a tummy ache if you forgot to take your enzymes with a meal? (Y)	86	n/a
Does a breakfast of toast and juice need more enzymes than a breakfast of bacon and egg? (N)	67	n/a
Do enzymes digest food? (Y)	n/a [b]	87
Are more enzymes needed to digest fat than carbohydrate? (Y)	n/a	94
Does a lunch of vegemite sandwiches need the same number of enzymes as a lunch of sausage rolls? (N)	n/a	80
Is it best to take enzymes before **and** during a meal? (Y)	63	76

(continued next page)

Vitamins

Do children who take enzymes need vitamin supplements? (Y)	93	n/a
Does malabsorption cause children with CF to need more vitamins than children without CF? (Y)	91	n/a

Malabsorption

Is firm poo a sign of possible malabsorption? (N)	93	n/a
Is very bad smelling poo a sign of possible malabsorption? (Y)	98	n/a
Is oily/greasy poo a sign of possible malabsorption? (Y)	96	n/a
Is a bloated tummy a sign of possible malabsorption? (Y)	91	n/a
Is less poo a sign of possible malabsorption? (Y)	60	n/a
Is pale, light brown or yellow poo a sign of possible malabsorption? (Y)	87	n/a
Are tummy cramps or pains a sign of possible malabsorption? (Y)	91	n/a
Is loose poo a sign of possible malabsorption? (Y)	93	n/a
Is passing poo more often each day a sign of possible malabsorption? (Y)	73	n/a

Fat

Which has the most fat - a boiled egg or a banana? (egg)	67	74
Which has the most fat - a slice of toast or a slice of cheese? (cheese)	96	63
Which has the most fat - a bowl of jelly or a bowl of custard? (custard)	n/a	81
Which has the most fat - a bowl of canned fruit or a bowl of custard? (custard)	98	n/a
Which has the most fat - a glass of milk or a glass of juice? (milk)	100	84

Energy

Which has the most energy - a glass of lemonade or a glass of milk? (milk)	74	78
Which has the most energy - a cheese sandwich or a jam sandwich? (cheese)	70	74
Which has the most energy - a bucket of hot chips or a boiled potato? (chips)	33	62
Which has the most energy - a slice of tomato or a sausage? (sausage)	49	n/a
Which has the most energy - a slice of tomato or a slice of ham? (ham)	n/a	73
Does a child with CF need less energy? (N)	63	n/a
For a child with CF,		
would drinking milkshakes give them more energy? (Y)	54	n/a
would adding cream to fruit and vegetables help increase their energy intake? (Y)	42	76
would drinking water help increase their energy intake? (N)	12	62
would eating meals and between meal snacks help increase their energy intake? (Y)	56	96
would adding more margarine or butter to hot food help increase their energy intake? (Y)	54	82
would sprinkling cheese on vegetables help increase their energy intake? (Y)	58	84
Do foods with a lot of fat in them have more energy than foods with a lot of carbohydrate? (Y)	n/a	42

Salt

Should children with CF stop eating salty foods? (N)	79	n/a
Do children with CF sweat more than children who do not have CF? (N)	19	n/a
Is the sweat of children with CF more salty than the sweat of children who do not have CF? (Y)	70	n/a
Which of the following food and drinks have lots of salt in them? Does - lemonade? (N)	54	73
cheese? (Y)	26	80
packet soup? (Y)	63	96
yoghurt? (N)	63	80
pizza? (Y)	70	98
Which has the most salt - a packet of Twisties or a bag of Jelly Beans? (Twisties)	74	98
Which has the most salt - a sausage or a slice of bread? (sausage)	65	n/a
Which has the most salt - a slice of ham or a slice of bread? (ham)	n/a	91
Which has the most salt - a bowl of custard or a bowl of soup? (soup)	63	100
Which has the most salt - a slice of tomato or a slice of cheese? (cheese)	65	86

[a] Correct response shown in parathenses, Y=Yes, N=No

[b] Not applicable. Questions which did not appear in the final questionnaires as they were altered or deleted after reliability testing

Analysis of the individual questions revealed that most children correctly answered a majority of the questions about enzymes. Fewer children were able to correctly identify which of two breakfasts (containing differing amounts of fat) required more enzymes with 9 of the 42 children indicating they did not know the answer to the question.

Three of the four questions asking which of two foods has the most fat were correctly answered by at least 32 of the 42 children. However, 14 of the children did not know that a slice of cheese contains more fat than a slice of toast.

Children did less well with the questions in the salt domain. Seven of the eleven questions were incorrectly answered by at least one-third of the children. Only 11 of the 42 children knew that cheese contains lots of salt and 14 of the children thought that a slice of tomato contains more salt than a slice of cheese.

The energy content of food was the least understood domain. Seven of the 12 questions were answered incorrectly by 20 or more of the 42 children. Only 14 of the children knew that a bucket of hot chips contains more energy than a boiled potato. Almost half the children were unaware of the variety of strategies suitable for increasing the energy intake of a child with CF, such as drinking milkshakes and adding cream, margarine, butter and cheese to food.

Interviewers reported that younger children appeared to be guessing many of the answers and seemed to prefer providing 'yes' as the response. The average correct score for children aged 6 to 8 years was 54%, which was lower than the score of 72% for 9 to 12 year olds.

Knowledge Questionnaire - Carers

Pre-intervention, the carers mean knowledge score was 85%. Questions about energy were not as well answered as those regarding vitamins, fat, enzymes, salt and malabsorption (Table 5.2). Performance on individual questions in the knowledge questionnaire is shown in Table 5.2.

Analysis of the individual questions revealed that nearly all the carers were aware of what foods require enzymes and that more enzymes are needed to digest fat than carbohydrate.

Most carers identified correctly which of two foods contained the most fat, except that one-third answered incorrectly that a banana contained more fat than a boiled egg.

At least 50 of the 55 carers correctly identified soup, pizza, Twisties and ham as good sources of salt. However, 8 carers answered incorrectly that a slice of tomato contains more salt than a slice of cheese and 11 carers were unaware that cheese contains lots of salt.

Most carers knew that children with CF, who are on enzymes due to malabsorption, need vitamin supplementation. Most carers were also aware of the signs of possible malabsorption.

The energy content of food was the least understood domain in the carers' questionnaire. Between 12 and 18 of the 55 carers answered the four questions about which of two foods contains the most energy incorrectly. Twenty-three carers answered correctly that food high in fat contains more energy than food high in carbohydrate with 13 of the carers indicating they did not know the answer to this question.

Most carers were aware of strategies to increase the energy intake of children with CF. However, 21 of the 55 carers answered incorrectly that drinking water would help increase energy intake.

Self-Management Questionnaire - Children

Appendix 3.2 illustrates that the children's self-management section required subjects to respond to a list of options for specific scenarios which were both

appropriate and inappropriate. The children's mean appropriate and inappropriate self-management scores were 55% and 21%, respectively. This means that children responded to a correct option, or did not respond to an incorrect option, on average, 55% of the time and responded to an incorrect option or did not respond to a correct option, on average, 21% of the time. Prompting caused children to indicate appropriate actions three times more often than unprompted responses and inappropriate actions 10 times more often than unprompted responses.

Half the children in the study indicated that their stools over the few months prior to the interview were suggestive of malabsorption. These children reported that they informed carers about the possibility of malabsorption only 75% of the time, on average.

Analysis of the remaining scenario-based questions is shown in Table 5.3.

Table 5.3

Frequency of only appropriate responses, and number of appropriate and inappropriate or non-responses (unprompted and prompted responses combined), given to scenario-based self-management questions within each domain by children with cystic fibrosis (CF) aged 6 to 11 years (n=42)

Question domain	% of children who gave only appropriate responses	No. of appropriate responses	No. of inappropriate or non-responses
		Mode (range)	Mode (range)
1. Methods for checking height and weight	67	4 (1 to 5)	0 (0 to 2)
2. Reasons why children with CF may have difficulty putting on weight	55	4 (0 to 7)	0 (0 to 1)
3. Actions to take when teased about taking enzymes	26	3 (1 to 4)	1 (0 to 4)
4. Situations requiring extra salt	81	2 (0 to 3)	0 (0 to 1)
5. Enzyme dose alterations in response to tummy pains the day after eating fish and chips	69	1 (0 to 1)	0 (0 to 1)

Most children were aware of at least two appropriate methods for checking their growth and weight. Only 26 of the 42 children were aware of at least two reasons why children with CF may have difficulty putting on weight.

Approximately three-quarters of the children chose three appropriate actions for dealing with the situation of a child at school teasing, making fun or laughing at them taking enzymes. However, several inappropriate actions for dealing with this situation were also indicated.

Most respondents were aware of the need for extra salt for children with CF when they have been sweating due to sport or hot weather.

Eight of the 42 children indicated an excessive increase in the dose of enzymes for a child with CF who had tummy pains the day after he ate a meal of fish and chips. Five of the children said the number of capsules should stay the same or did not know what to do.

Self-Management Questionnaire - Carers

Appendix 3.3 illustrates that the carers' self-management section required subjects to respond to a list of options for specific scenarios which were both appropriate and inappropriate. The carers' mean appropriate and inappropriate self-management scores were 74% and 32%, respectively. This means that carers responded to a correct option or did not respond to an incorrect option, on average, 74% of the time and responded to an incorrect option or did not respond to a correct option, on average, 32% of the time. Prompting had less effect on the responses given by carers compared with those given by children. Prompting caused carers to indicate appropriate actions two times more often than unprompted responses, and inappropriate actions four times more often than unprompted responses.

Carers answered that children with CF would be able to: manage signs of possible malabsorption with no supervision at a mean age of 12 years (range 8 to 18 years), and with some supervision and advice at a mean age of 9 years (range 5 to 15 years);

and take pancreatic enzyme capsules with no supervision or reminders at a mean age of 10 years (range 5 to 22 years), and with some supervision and reminders at a mean age of 7 years (range 2 to 19 years).

Analysis of the remaining scenario-based questions is shown in Table 5.4. Most carers identified three appropriate actions they would take if their child had signs of more than usual malabsorption. Most carers also said they would increase the number of enzyme capsules. Whether they would do this with or without the guidance of CF clinic staff is not known from these questions.

At least 52 of the 55 carers were aware of three appropriate methods for checking their child's growth and weight. But six carers reported they were not confident about judging how well their child was growing by using a growth chart. At least 41 of the 55 carers were aware of seven causes for children with CF having difficulty putting on weight.

All carers were aware that children with CF need more salt than usual when they sweat playing sport and in hot weather.

Most carers identified at least five appropriate strategies for addressing the issue of a child with CF looking thin. All carers indicated they would increase the number of enzymes if the child appeared to be malabsorbing. Increasing the dose of enzymes was not always dependent on discussing the issue with CF clinic staff.

Fourteen of the 55 carers indicated an inappropriate amount of time (such as the same day or a week later), or gave no response, when asked how long it would take for extra enzyme capsules to minimise malabsorption.

The majority of carers chose small increments in enzyme dosage to address signs of malabsorption. However, 6 of the 55 carers indicated an excessive increase in the dose of enzymes for a child who had experienced abdominal pain the day after a meal of fish and chips.

Table 5.4

Frequency of only appropriate responses, and number of appropriate and inappropriate or non-responses (unprompted and prompted responses combined), given to scenario-based self-management questions by carers of children with cystic fibrosis (CF) aged 2 to 11 years (n=55)

Question domain	% of carers who gave only appropriate responses	No. of appropriate responses	No. of inappropriate or non-responses
		Mode (range)	Mode (range)
1. Actions if signs of possible malabsorption	91	5 (2 to 6)	0 (0 to1)
2. Methods for checking height and weight	58	5 (3 to 6)	0 (0 to 2)
3. Reasons why children with CF may have difficulty putting on weight	73	7 (3 to 8)	0 (0 to 1)
4. Situations requiring extra salt	100	2 (2 to 3)	0 (0 to 0)
5. Enzyme dose alterations in response to tummy pains the day after eating fish and chips	85	1 (0 to 1)	0 (0 to 1)
6. Strategies for addressing thinness	87	7 (5 to 8)	0 (0 to 1)
7. Enzyme dose alterations in response to signs of malabsorption	60	1 (0 to 1)	0 (0 to 1)
8. Length of time for extra enzymes to minimise malabsorption	73	1 (0 to 1)	0 (0 to 1)
9. Actions if malabsorption continues >1 week after increasing daily dose	34	4 (1 to 4)	1 (0 to 2)
10. Strategies to deal with enzyme refusal	2	6 (3 to 8)	4 (0 to 10)
11. Strategies to deal with food refusal	4	6 (3 to 8)	6 (0 to 10)
12. Ways to reinforce desirable meal behaviours	82	3 (0 to 4)	0 (0 to 1)

Most carers indicated two appropriate actions for dealing with a child who still had signs of more than usual malabsorption one week after taking extra enzyme capsules each day. However, 37 of the 55 carers indicated the inappropriate response of increasing the number of capsules further. Fortunately all these carers said they would discuss the issue with the CF clinic staff.

Carers' use of behaviour management strategies for enzyme and food refusal was assessed by asking what action they would take in such situations. Most carers indicated up to four appropriate strategies, but at least two-thirds of carers indicated the use of a total of seven inappropriate strategies, to deal with this problem. Only 23 carers were prepared to give their child the choice to either eat the meal or have nothing until the next snack or meal time.

If a child who usually refuses to eat his evening meal came to the table when called, and began eating, most carers indicated appropriate ways to reinforce the desired behaviour.

Associations Between Scores

Analysis of associations between children's and carers' scores and socioeconomic index revealed: statistically significant positive correlations between carers' and children's knowledge (Pearson's r=0.32, p=0.036) and children's knowledge and appropriate self-management scores (Spearman's r=0.41, p=0.008); a statistically significant negative correlation between the carers' knowledge and inappropriate self-management scores (Spearman's r=-0.35, p=0.023); and no statistically significant associations between socioeconomic index and children's and carers' knowledge and self-management scores.

Longitudinal

Descriptive statistics for the changes that occurred as identified using the children's and carers' knowledge and self-management questionnaires are shown in Tables 5.5 to 5.7.

Table 5.5

Change in the nutrition and enzyme knowledge scores for children with cystic fibrosis aged 6 to 11 years and carers of 2 to 11 year olds

	Variable	Children maximum possible score=37		Carers maximum possible score=42	
		Intervention (Group 1) n=21	Control (Group 2) n=21[b]	Intervention (Group 1) n=27	Control (Group 2) n=28[c]
Pre-intervention	Mean score	23.3	23.7	35.4	36.0
	Standard error	1.1	1.1	0.92	0.6
	Mean percentage of maximum score (%)	63	64	84	86
Post-intervention	Mean score	27.5	24.4	37.2	37.1
	Standard error	1.1	1.1	0.6	0.6
	Mean percentage of maximum score (%)	74	66	88	88
12 month follow-up	Mean score	25.5	25.8	37.0	37.6
	Standard error	1.2	1.1	0.6	0.5
	Mean percentage of maximum score (%)	69	70	88	89
Post-minus pre-intervention	Score change	4.2	0.7	1.8	1.1
	Standard error	0.6	0.7	0.8	0.5
	Score difference[a] Group 1 - 2	3.5***		0.7	
12 month follow-up minus pre-intervention	Score change	2.2	2.1	1.6	1.6
	Standard error	1.1	0.9	0.8	0.5
	Score difference[a] Group 1 - 2	0.1		0.0	

[a] statistical significance from independent t-tests for between group changes
[b] except at 12 month follow-up, n=20
[c] except at 12 month follow-up, n=27
*** significant difference between groups, p=0.001

Table 5.6

Changes in the nutrition and enzyme appropriate self-management scores for children with cystic fibrosis aged 6 to 11 years and carers of 2 to 11 year olds (unprompted and prompted responses combined)

	Variable	Children maximum possible score=23		Carers maximum possible score=61	
		Intervention (Group 1) n=21	Control (Group 2) n=21[a]	Intervention (Group 1) n=27	Control (Group 2) n=28[b]
Pre-intervention	Mean score	12.5	13.0	45.3	45.0
	Standard error	0.7	0.8	0.7	0.7
	Mean percentage of maximum score (%)	54	57	74	74
Post-intervention	Mean score	13.7	13.1	47.2	46.8
	Standard error	0.7	0.7	0.5	0.5
	Mean percentage of maximum score (%)	59	57	77	77
12 month follow-up	Mean score	12.3	12.8	48.0	48.0
	Standard error	0.6	0.6	0.7	0.5
	Mean percentage of maximum score (%)	54	56	79	79
Post-minus pre-intervention	Score change	1.2	0.1	1.9	1.8
	Standard error	0.5	0.8	0.6	0.7
	Score difference Group 1 - 2	1.1		0.1	
12 month follow-up minus pre-intervention	Score change	-0.2	-0.2	2.7	3.0
	Standard error	0.7	0.9	0.5	0.7
	Score difference Group 1 - 2	0.0		-0.3	

[a] except at 12 month follow-up, n=20
[b] except at 12 month follow-up, n=27

Table 5.7

Changes in the nutrition and enzyme inappropriate self-management scores for children with CF aged 6 to 11 years and carers of 2 to 11 year olds (unprompted and prompted responses combined)

		Children maximum possible score=11		Carers maximum possible score=41	
	Variable	Intervention (Group 1) n=21	Control (Group 2) n=21[a]	Intervention (Group 1) n=27	Control (Group 2) n=28[b]
Pre-intervention	Mean score	2.4	2.3	12.9	13.5
	Standard error	0.4	0.2	1.0	0.9
	Mean percentage of maximum score (%)	22	21	32	33
Post-intervention	Mean score	2.7	1.9	13.7	13.1
	Standard error	0.3	0.2	0.1	1.0
	Mean percentage of maximum score (%)	24	18	34	32
12 month follow-up	Mean score	2.3	1.9	12.6	13.4
	Standard error	0.4	0.3	1.2	0.9
	Mean percentage of maximum score (%)	21	18	31	33
Post-minus pre-intervention	Score change	0.3	-0.4	0.8	-0.4
	Standard error	0.3	0.3	0.5	0.9
	Score difference Group 1 - 2	0.7		1.2	
12 month follow-up minus pre-intervention	Score change	-0.1	-0.4	-0.3	-0.1
	Standard error	0.3	0.3	0.8	1.0
	Score difference Group 1 - 2	0.3		-0.2	

[a] except at 12 month follow-up, n=20
[b] except at 12 month follow-up, n=27

The change in children's knowledge from pre- to post-intervention was statistically significantly different between the intervention and control groups (p=0.001) (Table 5.5). This difference in the change between the groups was 3.5 units in favour of the intervention group (where the maximum knowledge score was 37). The score for the intervention group changed from a score of 23.3 to 27.5 units and for the control group changed from 23.7 to 24.4. The score improvement from pre- to post-intervention of 4.2 units by the intervention group was statistically significantly different from the improvement of 0.7 units by the control group (p=0.001). This improvement in knowledge of the children in the intervention group occurred in all four domains, with the greatest improvement being with the fat and enzyme questions.

From post-intervention to 12 month follow-up, the knowledge scores of the children in the intervention group decreased and the control group's improved to a point where the mean knowledge score for each group differed by only 0.3 of a unit at 12 month follow-up. As such, the improvement in children's knowledge from pre-intervention to 12 month follow-up was only 2.2 units and the score difference of 0.1 units was not statistically significantly different between the groups (Table 5.5).

Overall, there were no statistically significant differences in carers' knowledge between groups before or after the intervention (Table 5.5). Table 5.5 shows that carers had a high baseline level of knowledge. The slight improvement in carers' knowledge in the intervention group was in the enzyme domain.

Tables 5.6 and 5.7 show that the children's and carers' self-management scores were similar between groups at each of the three time points measured, and the scores of each group were similar at each of the three time points. The slight differences between groups were not statistically significant. Also, there was no overall intervention or time effect noted when individual questions in the self-management questionnaires were analysed.

A separate set of questions in the validated self-management questionnaire assessed children's reported communication with carers when they experienced signs of

possible malabsorption (abdominal pain and loose, fatty, frothy or pale bowel actions). Post-intervention, a greater percentage of children in the intervention group compared to the control group reported communicating with their carers when they experienced signs of possible malabsorption (intervention group: pre-intervention 43% & post-intervention 57%; control group: pre-intervention 67% & post-intervention 48%). The t-test indicated that the statistically significant difference between the groups in favour of the intervention group continued to the 12 month follow-up time point (p=0.002), even though less children in both groups reported communicating with their carers when they experienced signs of possible malabsorption.

A positive outcome from the intervention for carers was the increase in the percentage of respondents in the intervention group indicating appropriate management for malabsorption, growth, salt supplementation, enzyme therapy and co-operation with meal times, immediately post-intervention.

Pre-intervention, the percentage of carers in the intervention group selecting only appropriate responses for what carers could do to check the growth and weight of children was greater in the control group, but the difference between groups post-intervention was not statistically significant.

Analysis of children's and carers' scores using Spearman's correlation revealed that the knowledge of the children in each group post-intervention and at 12 month follow-up was statistically significantly associated with their appropriate self-management score (post-intervention: intervention group: r=0.52, p=0.02, control group: r=0.46, p=0.03; 12 month follow-up: intervention group: r=0.53, p=0.01, control group: r=0.71, p=0.00).

Chi-square analysis, based on a contingency table, indicated that the difference in the distribution of the socioeconomic index for the intervention and control groups was not statistically significant. Also, one-way analysis of variance indicated that the difference between the socioeconomic index categories and the children's and carers' mean knowledge and self-management scores were not statistically significant. This

non-significant difference persisted regardless of whether or not intervention and control status were accounted for.

Discussion

The cross-sectional, pre-intervention results indicated that a high proportion of children and carers (34 out of 42 and 50 out of 55, on average, respectively) were familiar with when and how to administer pancreatic enzymes to ensure maximum activity, the consequences of inadequate enzyme therapy and the need to take a larger dose with food high in fat. However, it appeared that the respondents' knowledge about the fat content of food was insufficient, in that many were unable to identify which of two meals, containing significantly different amounts of fat, would need more enzymes. Similarly, McCabe (91) found that although children knew more enzymes are needed with high fat food, they took their usual enzyme dose with meals and snacks which contained varying amounts of fat. These results highlight the need for interventions to improve children's knowledge about the fat content of food and drinks. An increase in knowledge is likely to have a two-fold benefit, in that children may be more able to achieve the high fat diet recommended for individuals who have CF and also adjust their enzyme dosage in accordance with the amount of dietary fat consumed. An enhancement in the nutrition knowledge of children with CF and their carers would make them more capable of matching pancreatic enzyme doses to the amount of fat at each meal and snack, as recommended by the Australian PERT Guidelines (67, 68).

The longitudinal study indicated that the *Go and Grow with CF* nutrition and behaviour intervention program was not effective in increasing children's and carers' knowledge and self-management in the long-term. However, the intervention was effective in increasing children's knowledge in the short-term, particularly regarding fat and enzymes, irrespective of a family's socioeconomic index. It is difficult to determine whether the mean improvement from pre- to post-intervention of 11% in the knowledge score of the children's intervention group (representing 3.5 out of 37 units) was clinically significant, given that 30% of questions were answered incorrectly at the 12 month follow-up and that there was no improvement in any of

the other parameters assessed (as will be presented in the sections which follow). All the questions in the questionnaires were considered by the investigators to be important concepts to be understood by children with CF in order for them to be capable of optimising their nutritional intake. The improved mean post-intervention score of 74% may still be insufficient for achieving CF dietary and PERT recommendations most of the time. An accurate assessment of dietary intake, in conjunction with knowledge, would be advantageous in determining what level of knowledge is necessary for optimising nutritional status.

The knowledge scores of the children's intervention and control groups were similar to each other 12 months after the intervention program. From post-intervention to the 12 month follow-up, the knowledge of the intervention group decreased while the knowledge of the control group increased. The knowledge of the control group may have increased as a consequence of respondents actively seeking the correct answer to questions of which they were unsure of during a previous administration of the questionnaire. The decrease in the knowledge of the intervention group at the 12 month follow-up suggests that periodic nutrition and PERT education may be necessary to help children and carers both retain knowledge gains achieved by participating in the *Go and Grow with CF* program, and make further advancements. A variety of age-appropriate materials and programs may need to be implemented on a rotational basis in order to maintain interest in dietary and PERT information.

The short-term improvement in knowledge about fat and enzymes from participation in the pilot program suggests that the 1998 revised version of *Go and Grow with CF* (detailed in Chapter 6), which includes a fat target (12) and the Australian PERT Guidelines (67, 68), could assist families in knowing more about the energy and pancreatic enzyme needs of a child who has CF. Although knowledge does not equate with behaviour, there is potential for *Go and Grow with CF* to be effective in teaching families the what and how of achieving important nutrition recommendations for children who have CF.

Use of the questionnaires showed that the concept of food and drinks having energy (kilojoule) value was poorly understood by both children and carers. A transition

statement was included in the final questionnaire as poor understanding of the energy concept was detected during piloting. However, it appears that the transition statement did not adequately enlighten respondents, such that they appeared to think that the term energy referred to levels of activity. This is illustrated by many children, and over a third of carers, indicating incorrectly that a person's energy intake could be increased by drinking water. These results indicate the need to refine this section of the questionnaires, possibly by ending relevant questions with "… increase the amount of energy/kilojoules/calories you have eaten or drunk", rather than "… increase your energy intake", and by further alterations to the transition statement, in order to improve the respondent's understanding about the energy concept.

Most children and carers were aware of the relationship between inadequate dietary intake and poor gains in weight and height. Carers were more aware than children of other causes of poor growth, such as excessive faecal losses and increased requirements. Most carers, but only half the children, indicated energy boosters that could be used to help increase a child's energy intake. Most respondents were aware they could monitor the effects of dietary interventions using growth charts, but a small percentage of carers indicated they were not confident in judging their child's progress using this tool.

Children's knowledge about the salt content of food was average. It is likely they had difficulty distinguishing between the different taste sensations, as many indicated that several sweet foods have a high salt content. Children and carers knew that some commonly eaten foods are high in salt, such as pizza, soup and sausages. However, it was surprising to find many children and carers did not know the nutritional value of cheese. Cheese is a convenient source of fat and salt in addition to being a valuable source of protein and calcium. Improved knowledge about the value of cheese could cause an increase in the level consumed and could make a significant contribution toward children with CF achieving their elevated dietary requirements.

A question was included in the self-management questionnaire to assess if respondents could indicate situations when extra salt would be required. Prior to prompting, this question was misinterpreted by some respondents who replied that extra salt is always required. Misinterpretation of the question could be overcome by a transition statement, such as "Children with CF need more salt than other children because they lose more through their skin. Tell me when a child with CF needs to take more salt than what they usually have".

Achieving and maintaining good nutritional status requires appropriate self-management of dietary intake and PERT (86). Although carers were knowledgeable about the signs of possible malabsorption, the cross-sectional study suggests that the problem is not always dealt with adequately. For example, the results indicated that children do not always inform carers about the symptoms of malabsorption, carers expect young children to manage malabsorption and the administration of pancreatic enzymes without supervision and doses are often increased excessively in an attempt to alleviate signs of possible malabsorption. These findings illustrate the need for children to clearly understand the purpose of, and dosing methods for, pancreatic enzyme therapy and that carers need to be encouraged to co-manage this aspect of their child's CF treatment more closely.

The importance of carers, children and health-care providers communicating and managing malabsorption together is reinforced by the finding that some children were unable to suggest appropriate changes a child with CF could make to his pancreatic enzyme dose if he experienced malabsorption the day after a very high fat meal. Another concern is that some children and carers chose excessive increases in pancreatic enzyme doses to alleviate possible malabsorption, and expected an unrealistic immediate response, or did not know how long it would possibly take for the problem to be resolved. Carers indicated they would increase the dose further if there was no improvement in absorption and many did not indicate they would discuss this with CF clinic staff. The incidence of fibrosing colonopathy amongst children on high doses of lipase reinforces the importance of ensuring that excessive pancreatic enzyme dosing is avoided (69, 151). PERT guidelines suggest an upper daily limit be established for each patient and the Australian guidelines recommend

assessment of enzyme administration, storage and compliance, the need for further investigation and the use of adjunctive therapies rather than allowing the dose to rise without resolution of the symptoms (67).

The cross-sectional study showed that children were limited in their ability to suggest several appropriate strategies for dealing with being teased for taking enzymes. In addition to appropriate actions, over half the children also indicated they would get upset and some suggested inappropriate actions, such as reciprocal teasing, hitting the offender or stopping and hiding when taking enzymes. It may be possible to improve this apparent lack of confidence about taking pancreatic enzymes at school by increasing children's level of knowledge and helping them to explore suitable responses to being teased (115).

Carers also indicated the use of both appropriate and inappropriate strategies on the same occasion for dealing with a child's refusal of enzymes or food. Although routine prompting during the interview may have caused over-reporting of the use of inappropriate strategies, the results supported the need for education programs for carers of children with CF to include a component on behaviour management. Carers need to be confident about the appropriateness of ignoring enzyme and food refusal, even at the risk of a meal or snack being omitted, in order to achieve adherence to treatment in the long-term (124).

The longitudinal study indicated that the proportion of carers in the intervention group indicating appropriate management of several aspects of nutrition and PERT was significantly different from the proportion of carers in the control group immediately post-intervention, although this result was not duplicated at the 12 month follow-up time point. Twelve months after the intervention, knowledge and self-management of both the children's and carers' control group also improved and the intervention group declined, such that the scores of the groups were similar. The improvement in the children's and carers' control groups may be a consequence of the respondents actively seeking information about issues they felt they did not answer correctly each time they were interviewed. This positive effect of the study provides further evidence of the need for, and value of, continuing education.

Improvement in knowledge does not guarantee positive changes in dietary behaviour. But, the positive association between knowledge and appropriate self-management post-intervention and at the 12 month follow-up time point, for both the children's intervention and control groups, concurs with social learning theory constructs which maintain that an individual's ability to perform certain behaviours depends on the acquisition of adequate knowledge (115, 129). Children's responses to questions about reporting signs of possible malabsorption suggested that there was both a short- and long-term improvement in the intervention group's communication with carers about problems associated with PERT. The improvement in communication about possible malabsorption is an important outcome of the program and possibly indicates that *Go and Grow with CF* equipped families with information to assess and discuss an aspect of the child's gastrointestinal management, which may in turn improve clinical outcomes.

5.4 Behaviour

Methods (Section 4.5)

Results

Data available for analysis of the BPFAS included all children enrolled in the clinical trial, being 58 two to 11 year olds pre- and post-intervention and 57 children at the 12 month follow-up time point.

Descriptive statistics for the BPFAS questionnaire, as reported by carers, are shown in Tables 5.8 to 5.11.

Table 5.8

Frequency and problem scores for child behaviour and carers' responses, as reported by carers of children with cystic fibrosis aged 2 to 11 years immediately prior to the intervention, using the Behavioural Pediatrics Feeding Assessment Scale (106)

	Intervention group n=29	Control group n=29	Estimate (95% CI)[a]	P-value[b]
	mean±sd (range)	mean±sd (range)		
Child behaviour - frequency max. possible score = 125	48.2±10.0 (33 to 66)	53.3±10.5 (33 to 79)	-5.1 (-10.5, 0.3)	0.06
Child behaviour - problems max. possible score = 25	3.1±3.7 (0 to 14)	4.5±4.8 (0 to 18)	-1.4 (-3.7, 0.8)	0.21
Carer's feelings/strategies - frequency max. possible score = 50	16.4±4.6 (10 to 29)	19.3±6.2 (10 to 36)	-2.9 (-5.8, -0.1)	0.046
Carer's feelings/strategies - problems max. possible score = 10	1.0±1.6 (0 to 7)	2.0±2.8 (0 to 10)	-1.0 (-2.2, 0.18)	0.28

[a] estimate (confidence interval) for the difference between groups
[b] independent t-test for the difference between groups

Table 5.9

Frequency and problem scores for child behaviour and carers' responses, as reported by carers of children with cystic fibrosis aged 2 to 6.9 years and 7 to 11 years immediately prior to the intervention, using the Behavioural Pediatrics Feeding Assessment Scale (106)

	Children 2 to 6.9 years n=31	Children 7 to 11 years n=27	Estimate (95% CI)[a]	P-value[b]
	mean±sd (range)	mean±sd (range)		
Child behaviour - frequency max. possible score = 125	54.2±8.0 (36 to 70)	46.7±11.6 (33 to 79)	7.5 (2.3, 12.7)	0.005
Child behaviour - problems max. possible score = 25	4.4±4.1 (0 to 17)	3.2±4.5 (0 to 18)	1.2 (-1.1, 3.4)	0.30
Carer's feelings/strategies - frequency max. possible score = 50	18.7±4.6 (11 to 29)	16.9±6.6 (10 to 36)	1.8 (-1.1, 4.8)	0.21
Carer's feelings/strategies - problems max. possible score = 10	1.6±2.3 (0 to 8)	1.4±2.3 (0 to 10)	0.18 (-1.0, 1.4)	0.77

[a] estimate (confidence interval) for the difference between age groups
[b] independent t-test for the difference between age groups

Table 5.10

Changes in frequency scores for child behaviour and carers' responses, as reported by carers of children with cystic fibrosis aged 2 to 11 years, using the Behavioural Pediatrics Feeding Assessment Scale (106)

		Child frequency maximum possible score = 125		Carer's frequency maximum possible score = 50	
		Intervention (Group 1) n=29	Control (Group 2) n=29[c]	Intervention (Group 1) n=29	Control (Group 2) n=29[c]
Pre-intervention	Mean score	48.2	53.3	16.4	19.3
	Standard error	1.86	2.0	0.86	1.2
	(range)	(33 to 66)	(33 to 79)	(10 to 29)	(10 to 36)
	Mean percentage of maximum score (%)	39	43	33	39
Post-intervention	Mean score	45.1	49.4	15.3	18.5
	Standard error	1.7	2.0	0.8	1.1
	(range)	(31 to 66)	(33 to 77)	(10 to 24)	(10 to 29)
	Mean percentage of maximum score (%)	36	40	31	37
12 month follow-up	Mean score	43.7	47.5	15.1	18.0
	Standard error	1.5	1.7	0.8	1.0
	(range)	(29 to 59)	(30 to 68)	(10 to 27)	(10 to 32)
	Mean percentage of maximum score (%)	35	38	30	36
	Score change	-3.03	-3.83	-1.07	-0.79
	Standard error	1.14	1.49	0.81	0.70
Post-minus pre-intervention	Score difference Group 1 - 2	0.79		-0.28	
	P-value[a]	0.67		0.80	
	95% CI[b]	(-3.0, 4.5)		(-2.4, 1.9)	
	Score change	-4.48	-5.61	-1.31	-0.82
12 month follow-up minus pre-intervention	Standard error	1.29	1.19	1.00	0.63
	Score difference Group 1 - 2	1.12		-0.49	
	P-value[a]	0.52		0.68	
	95% CI[b]	(-2.4, 4.6)		(-2.9, 1.9)	

[a] independent t-tests for the difference between group changes

[b] confidence interval for the difference between group changes

[c] except at 12 month follow-up, n=28

Table 5.11

Changes in problem scores for child behaviour and carers' responses, as reported by carers of children with cystic fibrosis aged 2 to 11 years, using the Behavioural Pediatrics Feeding Assessment Scale (106)

		Child problem		Carer's problem	
		Intervention (Group 1) n=29	Control (Group 2) n=29[g]	Intervention (Group 1) n=29	Control (Group 2) n=29[g]
Pre-intervention	Rate of problems (%)[a]	66	76	48	66
	Logit of rate[b]	0.64	1.15	-0.07	0.64
	Logit difference Group 1 - 2	-0.50		-0.71	
	Standard error of the difference	0.58		0.54	
	P-value[c]	0.39		0.19	
Post-intervention	Rate of problems (%)	59	66	21	48
	Logit of rate	0.35	0.64	-1.34	-0.07
	Logit difference Group 1 - 2	-0.29		-1.27	
	Standard error of the difference	0.54		0.59	
	P-value	0.59		0.031	
12 month follow-up	Rate of problems (%)	55	68	24	43
	Logit of rate	0.21	0.78	-1.15	-0.26
	Logit difference Group 1 - 2	-0.57		-0.89	
	Standard error of the difference	0.55		0.58	
	P-value	0.30		0.12	

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		Child problem P-value	Carer's problem P-value
	Time[d]	0.50	0.013
Likelihood ratio test	Group[e]	0.17	0.004
	Group by Time[f]	0.94	0.47

[a] percentage of subjects with one or more problems
[b] if the rate is p then the logit of the rate is logit $(p) = \log(p/1-p)$
[c] analysis of the difference between groups based on a comparison of estimated
    co-efficients from the logistic regression model
[d] main effect of time on the rates (intervention and control group combined)
    H_0: $rate_2 = rate_3 = rate_5$
[e] main effect of group over all five time points, ie average group effect
    H_0: average difference between groups over all five time points = 0
[f] difference of the rates between groups over all five time points
    H_0: $\Delta rate_2 = \Delta rate_3 = \Delta rate_5$
[g] except at 12 month follow-up, n=28

Cross-sectional

Table 5.8 shows that prior to the intervention the frequency of carers' responses to child meal-time behaviours in the intervention group was statistically significantly lower than the control group (p=0.046). There were no statistically significant differences between the intervention and control groups for the child frequency and child and carers' problem scores (Table 5.8). There were also no statistically significant differences between genders of the children for the child and carer's frequency and problem scores immediately prior to the intervention. However, an assessment of age indicated that carers reported a statistically significantly higher rate of inappropriate child meal-time behaviours for children less than 7 years compared to those aged 7 or more years (p=0.005) (Table 5.9).

Although the frequency scores of inappropriate child behaviours averaged at 38%, carers reported low levels of problems (average of 15%) with child behaviours and carers' responses to dealing with dietary-related issues (mean of 3.8 out of 25 and 1.5

out of 10 problems with child behaviours and carers' responses, respectively, when the data of all subjects were combined).

Longitudinal

Changes in frequency scores for child behaviour and carers' responses from pre- to post-intervention and pre-intervention to 12 month follow-up were not statistically significantly different between the intervention and control groups (Table 5.10). Frequency scores for child behaviours and carers' responses decreased consecutively in both groups over these three data collection time points.

With regard to problems with child behaviours (ie, no problems or 1 or more problems) (Section 4.5), the rates at each time point were higher in the control group than the intervention group, but there were no significant differences between groups either over all time points or at any given time point, ie. as shown in Table 5.11, there was no evidence of independent time or group effects, or a group by time interaction for the child behaviour problems.

In contrast, the rates of problems with carers' responses were higher in the control group than the intervention group. The rates of problems with carers' responses reached statistical significance post-intervention (p=0.031) and when considered over all time points (Likelihood ratio test of group p=0.004) (Table 5.11). Table 5.11 shows that the time effect was statistically significant, due to the lower rates in both groups post-intervention and at the 12 month follow-up time point compared with pre-intervention (Likelihood ratio test of time p=0.013). However, there was no evidence of a group by time interaction.

Discussion

Prior to the intervention, there was a statistically significant difference between groups in the frequency of carers feelings/strategies for inappropriate child meal-time behaviours as measured by the BPFAS (p=0.046). There were no statistically significant differences in the scores obtained between genders of the children, but there was a statistically significant difference in the scores obtained between younger

and older children (Table 5.9). The significantly higher rate of inappropriate child behaviours in younger children (p=0.005) enrolled in the clinical trial supports the findings of Crist et al (106) who developed the questionnaire. The mean frequency and problem scores reported by Crist et al (106) were higher than the mean scores for all the children in our study, possibly because the children in the study by Crist et al (106) were younger, as other characteristics were similar (pulmonary function and %IBW status). It is likely that young children are more prone to exhibiting adverse meal-time behaviours than are older children (104). Inappropriate meal-time behaviours may cause energy intakes to be reduced (106), and thereby adversely affect the clinical status of children with CF.

Pre-intervention, most carers (74%) in our study reported that they either had no problems or only one out of the 10 possible problems with their responses to their child's food-related behaviours. This prompted an analysis based on the binary variable of whether or not they had problems. Given that seven of these 10 questions in the carers' section of the BPFAS have the potential to reflect carers' level of self-efficacy, the results suggest that carers had a high level of self-efficacy about the food-related behaviours of their child with CF. For example, if a carer reported that they got frustrated when feeding his/her child and then indicated that this was not a problem,' then this could be interpreted that this carer's level of self-efficacy regarding this issue was high. Nonetheless, the moderate frequency of adverse food-related behaviours of children with CF in our study supports the need for programs which teach carers to avoid strategies that may reinforce non-eating behaviour (coaxing) and guide carers to employ contingency praise and the setting of limits to manage unacceptable behaviour at meal-time. Stark et al (103) advocate that, regardless of the nutritional status of the child with CF, all parents may benefit from routine behavioural management training, in conjunction with nutrition education.

The effects of the *Go and Grow with CF* intervention program on the frequency scores for child behaviour and carers' responses, and the problem score for child behaviour, were not statistically significant. There were, however, statistically significant differences between the intervention and control group for the problem

scores for carers' responses to children's behaviour. Less carers in the intervention group endorsed items in the carers' responses section as problematic, immediately post-intervention (p=0.031) (Table 5.11). However, this statistically significant difference between groups in problem scores for carers' responses was not maintained 12 months after the intervention. The observation of fewer carers in the control group indicated problems at 12 month follow-up compared with the post-intervention time point may be a consequence of the children being one year older and, thus, exhibiting less adverse meal-time behaviours. There was no evidence of a group by time interaction (Table 5.11).

The lack of detection of a statistically significant effect of the intervention on dietary-related behaviours problems of children with CF 12 months after the intervention in this study may have been limited by a number of factors, including:

- low pre-intervention scores (an average over both the child and carer sections of the questionnaire of 40% and 15% for frequency and problem scores, respectively), suggesting low baseline levels of inappropriate behaviour

- carers in both groups actively seeking to address dietary-related issues brought to their attention after implementation of the questionnaire

- the intervention not specifically addressing the dietary-related issues assessed by the BPFAS

- the intervention was not sufficiently intensive to address the dietary-related issues assessed by the BPFAS, and

- children moving through a difficult age-related stage regardless of being in the intervention or control group.

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5.5 *Dietary intake*

Methods (Section 4.6)

Results

Data available for analysis of the FFQs included all 58 children at baseline, 57 of the children pre- and post-intervention, 55 of the children at 6 month follow-up and 56 children at the 12 month follow-up time point. Omissions were mostly due to carers being too busy to complete the questionnaires.

Cross-sectional

Descriptive statistics for the GPHNS FFQ show that there were no statistically significant differences in intakes for the nutrients assessed between the intervention and control group prior to the intervention (Table 5.12). The mean dietary intakes of energy, fat, protein, zinc and sodium of the children with CF, as measured using the GPHNS FFQ, were mostly in excess of recommended levels for both the normal population and individuals with CF (Table 5.12). Fat intake, expressed as a percentage of energy, was the only nutrient assessed as being lower than the recommended level for those with CF (being 1% and 3% lower than the recommended level for intervention and control group, respectively).

The energy intake of the females with CF in the study, expressed as a percentage of the age-, gender- and weight-adjusted RDI, was statistically significantly higher than males (p=0.050). Thirteen of the 27 females had energy intakes in excess of 200% of the RDI, in contrast to only 7 of the 30 males having energy intakes in excess of 200% of the RDI (Chi-square test p=0.093). There were no other statistically significant differences between the intakes of males and females for the nutrients assessed.

Table 5.12
Mean daily dietary intake of 2 to 11 year old children with cystic fibrosis immediately prior to the intervention, using a food frequency questionnaire (61, 64)

Nutrient	Unit	Recommended	Intervention group, n=29 mean±sd (range)	Control group, n=29 mean±sd (range)	Estimate (95% CI)[f]	P-value[g]
Energy	MJ		14.9±8.8 (6.8 to 43.7)	14.5±5.9 (6.7 to 30.1)	0.4 (-3.6, 4.5)	0.82
	%RDI[a]	120 to 150[b]	199±107 (87 to 550)	211±105 (84 to 545)	-12 (-68, 44)	0.68
Fat	g	>100[c]	159±109 (60 to 504)	147±68 (53 to 320)	12 (-36, 61)	0.61
	% MJ	40[d]	39±5 (28 to 47)	37±5 (23 to 45)	2 (-1, 4)	0.28
Protein	g		123±66 (57 to 321)	121±47 (55 to 252)	2 (-28, 33)	0.88
	% MJ		14±2 (12 to 19)	14±2 (12 to 18)	0 (-1, 1)	0.77
Zinc	mg	4.5 to 9[e]	17±10 (7 to 46)	17±8 (8 to 42)	0 (-5, 5)	0.98
Sodium	g	0.3 to 2.3[e]	1.9±3.7 (1.6 to 16.9)	4.7±2.2 (2.1 to 10.7)	0.8 (-1.4, 1.8)	0.82

[a] percentage of the recommended daily intake for age, gender and weight
[b] for individuals with CF (26, 27)
[c] for individuals with CF (12)
[d] for individuals with CF (74)
[e] for 2 to 11 year olds in the general population (72)
[f] estimate (confidence interval) for the difference between groups
[g] independent t-test for the difference between groups

Table 5.13
Multivariate analysis of dietary intake measurements of 2 to 11 year old children with cystic fibrosis in the intervention group (n=27) and control group (n=26), using a food frequency questionnaire (61)

Measure			F-ratio (d.f.)[f]	P-value
Energy	MJ	Time[c]	1.48 (4, 48)	0.22
		Group[d]	0.76 (1, 51)	0.39
		Group*Time[e]	0.61 (4, 48)	0.66
	%RDI[a]	Time	1.29 (4, 48)	0.29
		Group	0.01 (1, 51)	0.96
		Group*Time	1.00 (4, 48)	0.42
Fat	g	Time	1.38 (4, 48)	0.26
		Group	0.99 (1, 51)	0.32
		Group*Time	0.56 (4, 48)	0.70
	% MJ[b]	Time	0.82 (4, 48)	0.52
		Group	1.81 (1, 51)	0.32
		Group*Time	0.98 (4, 48)	0.42
Protein	g	Time	1.01 (4, 48)	0.41
		Group	0.46 (1, 51)	0.50
		Group*Time	0.69 (4, 48)	0.60
	% MJ[b]	Time	0.48 (4, 48)	0.75
		Group	1.71 (1, 51)	0.20
		Group*Time	1.68 (4, 48)	0.17
Zinc	mg	Time	0.39 (4, 48)	0.81
		Group	0.26 (1, 51)	0.61
		Group*Time	0.39 (4, 48)	0.82
Sodium	g	Time	0.82 (4, 48)	0.52
		Group	0.58 (1, 51)	0.45
		Group*Time	0.14 (4, 48)	0.97

[a] percentage of the recommended daily energy intake for age, gender and weight
[b] energy contribution as a percentage of total energy intake
[c] main effect of time on the measure (intervention and control group combined)
    H_0: $\mu_1 = \mu_2 = \mu_3 = \mu_4 = \mu_5$
    Wilks' Lambda multivariate test
[d] main effect of group over all five time points, ie average group effect
    H_0: average difference between groups over all five time points = 0
    Repeated measures analysis of variance
[e] difference of the measure between groups over all five time points
    H_0: $\Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$
    Wilks' Lambda multivariate test
[f] numerator degrees of freedom, denominator/error degrees of freedom

Longitudinal

Multivariate analysis indicated that the effects of group, time and group by time were all non-significant for the measurements of dietary intake, regardless of adjustment for covariates. The results are summarised in Table 5.13.

Discussion

As measured by the GPHNS FFQ, there was no statistically significant time trend, difference between groups or group by time interaction in the nutrient intakes of children with CF in this clinical trial (Table 5.13). *Go and Grow with CF* does not appear to have had an effect on dietary intake.

There is the possibility that the dietary intakes of the children with CF in this study were grossly over-estimated using the GPHNS FFQ (Table 5.12). The reported intakes of children with CF in this study were considerably higher than the theoretical requirements for CF (12, 26, 27, 74) and non-CF populations (72) (Table 5.12) and the intakes of children with CF reported in numerous dietary studies, using methodologies other than FFQs, which were close to age- and gender-specific RDIs (see Inadequate intake of Section 2.4). Also, the possibility of extremely high energy intakes is not supported by the subjects' sub-optimal growth parameters and mild to moderate deterioration in lung function. Table 5.1a shows that the pre-intervention mean height and weight of the children in the study were below normal and that the mean values for the parameters of lung function were mostly in the normal range.

Although the most common bias with dietary intake studies is considered to be under-reporting (134), it is not surprising that the results appear to be an over-estimation, given that the emphasis placed on individuals with CF to consume a diet high in energy, fat, and salt may have caused carers to over-report the dietary intake of their child. For example, carers of one-third of the subjects reported intakes in excess of 200% of the RDI for energy. However, it is not possible to conclusively state that the dietary intakes obtained using a FFQ with carers of children with CF in this study are truly an overestimation of actual intake, without comparison to a superior method of dietary assessment, or estimations of energy expenditure and

losses, as comparisons with RDIs are limited by differences in dietary intake methodology (Section 2.4).

The possibility of FFQs over-estimating the dietary intake of children with CF is supported by several other studies which have used FFQs with children and adults who do not have CF (62-64, 152-154). The comparison by Wheeler et al (64) between two FFQs (including the GPHNS FFQ used in this study) and 12-day weighed food records completed by adults (on behalf of themselves) indicated that differences between the two FFQs were small, but the differences between the FFQs and weighed records were large. In particular, FFQ intakes were significantly higher than weighed record intakes for nutrients largely derived from fruit and vegetables (sugars, fibre, potassium, magnesium, B-carotene and vitamin C). The study by Jenner et al (63), which involved 11 to 12 year old children completing a 175-item based FFQ and 14 24-hour food records of their intake, indicated that mean nutrient intakes obtained using the FFQ were consistently higher than those reported by the diet records. Thirdly, results obtained in a study by Garnett et al (62), in which 12 to 18 year old adolescents with insulin dependent diabetes mellitus completed a FFQ, also indicated intakes higher than those obtained from 4-day estimated food records and higher than intakes of an Australian reference group of 12 to 15 year old children who used a 24-hour food diary (Department of Community Services and Health 1988).

It is not possible to determine a correction factor by which FFQs under- or over-estimate energy and nutrient intakes as the process is complicated by differences between studies in subject characteristics, questionnaire design (list- or meal-based, number and types of foods listed, frequency categories, portion sizes), comparative tools (weighed food records, 24-hour recalls), duration of the record keeping periods, order of administration of the tools, nutrients reported and nutrient data bases used. These differences cause a great deal of variation in the correlation coefficients of nutrient intakes within and between studies assessing the validity of FFQs. It is possible that the variation in correlation coefficients is also influenced by the degree

of under-estimation of usual dietary intake by weighed and estimated food records (154, 155).

Factors which may have caused dietary intakes in this study to be over-estimated are numerous and are listed below.

1. Memory errors regarding the frequency of consumption and the sizes of food portions consumed.

2. Errors in determining appropriate frequency categories and serve sizes (156), particularly for items in mixed dishes.

3. Respondent fatigue due to the self-administered questionnaire containing more than 150 items (157).

4. Misclassification during coding.

5. Differences between actual intake and nutrient databases (due to seasonal changes, storage and differences in cooking methods).

6. Non-validation of the GPHNS FFQ for children with CF, due to low availability of subjects for the validation process, as most eligible children were enrolled in the study, subject burden and time constraints.

7. Carers' difficulty in distinguishing between what their children actually ate and what they would like their child with CF to have consumed (in order to meet elevated CF requirements).

8. Carers' estimating what school-aged children consumed over several hours most days of the week when the child was at school.

9. The majority of the carers who completed the FFQs being females (mothers of the children). Females have been observed to be more likely than males to overestimate nutrient intakes using FFQs (62, 64).

10. Carers responses being influenced by what they thought the investigator/dietitian would expect the children to consume.

The validity of parental reports of the dietary intake of children, using any of the variety of dietary recording and reporting methods available, is not well established. Jenner et al (63) noted differences between the nutrient intakes obtained from 11 to 12 year old children and parents (on behalf of the children) using the same FFQ. In

contrast, Byers et al (158) found parents' reports of their children's dietary intake of nutrients derived from fruits and vegetables, using a 111-item FFQ, to be reasonably correlated with blood levels of carotene and vitamin C.

The bias that seems to have been introduced by carers completing the FFQs on behalf of the children in our study, render the cross-sectional data as unsuitable for comparison with other CF dietary studies. Rockett and Colditz (58) recently concluded in a review of dietary intake methods used with children and adolescents that the principal advantage of FFQs is in ranking the diet of individuals, not in quantifying individual intake. Theoretically, the possible overestimation of dietary intakes should not have precluded the detection of within-person change in the longitudinal study as the same FFQ was administered to both the intervention and control group under the same conditions during all five data collection periods. The validity of FFQs for longitudinal studies has not yet, to the author's knowledge, been established. Alternative methods of dietary assessment, that have minimal subject burden, may need to be considered for future longitudinal studies involving children with CF. Rockett and Colditz (58) suggest the use of the internet and videoconferencing to directly connect researchers to their subjects and small portable tape recorders embedded with computer chips.

As this research project was an assessment of a pilot intervention program and families of children with CF are faced with many demands on their time, minimal subject burden regarding data collection was a primary objective. The selection of a tool to assess dietary intake during the clinical trial was limited by the lack of assessment of dietary tools for use with children. The literature supported the use of a FFQ in that FFQs can be used to obtain an estimate of the mean intake of nutrients for a group (60), while being less time consuming than other methods. Minimal subject burden was important for maximising both participation in the intervention and completion rates of the FFQs. Wheeler et al (61) reported that FFQs administered at intervals of at least 3 months (which was the minimum data collection time interval during this study) are characterised by high completion rates. Use of a superior method, namely weighed dietary records (which are considered the

gold standard for determining nutrient intakes), may have seriously limited subject recruitment and retention. There is also the possibility that previous exposure to food records, through faecal fat balance studies (Section 2.5), would affect the data obtained if this method had been used as carers may alter the child's usual intake to what they remember as being easier to record.

5.6 Body composition

Methods (Section 4.7)

Results

Of the children aged 2 to 11 years, height and weight data available for analysis included 58 subjects at baseline and pre-intervention and 57 subjects post-intervention and at the 6 and 12 month follow-up time points. Of the children aged 5 to 11 years, skinfold and girth data available for analysis included 39 subjects at baseline, pre- and post-intervention, and 38 subjects at the 6 and 12 month follow-up time points.

At baseline, pre- and post-intervention, all subjects were pre-pubertal. At 6 months follow-up, three females were assessed as being at Tanner Stage 2. At 12 months follow-up, one of these three females had progressed to Tanner Stage 3 and a total of five females and one male were assessed as being at Tanner Stage 2. Also, at the 12 month follow-up time point one female subject had been on insulin for 3 months due to diagnosis of CF-related diabetes and one male subject had been receiving growth hormone for five months due to inadequate bone growth.

Data from two children were excluded from the cross-sectional analysis only due to the extremes represented by the excessive body weight of one child (127% IBW) and the inadequate bone growth of the other (z-height -3.61 and bone age 3 years less than chronological age). When these two children were compared with subjects in the study of the same gender, the next highest %IBW value was substantially lower at 112%-and the next lowest z-height score was nearly two units higher at -0.96.

Cross-sectional

Height and weight

Table 5.1a shows that the height and weight z-scores of the children in the intervention and control group were not statistically significantly different prior to the intervention.

Table 5.14 shows that the mean height and weight z-scores of both the males and females aged 2 to 11 years immediately prior to the intervention were less than zero, indicating that the children with CF were shorter and lighter for age and gender than the NCHS reference population. If the children with CF were similar to the NCHS reference population then the height and weight z-scores would have been closer to zero. The difference in the height of the children with CF compared to the NCHS reference group persisted when parental height was taken into consideration (Table 5.14), suggesting that the subjects' shorter stature cannot be attributed to a familial predisposition. If the parents of the children with CF were short then the height-adjusted z-scores would have been closer to zero than the child's height z-score. Table 5.14 also shows that the mean weight and height z-scores for females were significantly lower than those for males ($p < 0.05$).

The mean %IBW value was 99.0%, indicating that the subjects' weights were similar to their nominal ideal weight for height and suggesting that the children with CF in the study were adequately nourished (acceptable range 90 to 110% (18)). Table 5.14 shows that the %IBW values were not statistically significantly different between genders. It should be noted, however, that three of the 27 females had %IBW values below the acceptable lower bound of 90% for children with CF. The common characteristics between these three females were that their ages ranged between 9.5 and 11.5 years and they were rated as being Tanner pubertal stage 2 when the post-intervention measurements were taken a few months after the pre-intervention data collection period. It may be that these females had experienced a rapid gain in height related to the onset of puberty around the time that the pre-intervention measurements were taken.

The heights and weights and anthropometric measures of the children with CF aged 6 to 11 years were also compared to the COGRO reference group (Table 5.15). Similar to comparisons with the NCHS reference group (Table 5.14), the children with CF were significantly shorter and lighter than the COGRO reference group (p<0.05) (Table 5.15).

Table 5.14

Height and weight of 2 to 11 year old pre-pubertal children with cystic fibrosis immediately prior to the intervention

Measure	Females n=27	Males n=27	Estimate (95% CI)[d]	P-value[e]
	mean±sd (range)	mean±sd (range)		
z-weight[a]	-0.9±0.7 (-2.5 to 0.5)	-0.3±0.9 (-2.3 to 1.3)	-0.6 (0.18, 1.03)	0.007
z-height[a]	-0.8±1.0 (-2.9 to 1.4)	-0.2±0.9 (-2.1 to 1.5)	-0.6 (0.09, 1.09)	0.021
z-height adjusted for parental height[b]	-0.7±0.9 (-2.9 to 1.0)	-0.4±0.9 (-2.3 to 1.2)	-0.3 (-0.17, 0.83)	0.20
%IBW[c]	99.0±9.7 (79.1 to 119.2)	99.0±7.4 (90.0 to 115.0)	0.0 (-4.69, 4.72)	0.99

[a] z-score standardised for age and gender (136)
[b] (137)

[c] percent ideal body weight (18)
[d] estimate (confidence interval) for the difference between females and males
[e] independent t-test for the difference between females and males

Table 5.15

Anthropometry of 6 to 11 year old pre-pubertal children with cystic fibrosis (CF) and the COGRO reference group (143)

Measure	Females				Males			
	CF n=17 mean±sd	Reference n=145 mean±sd	95% CI[e]	P-value[f]	CF n=14 mean±sd	Reference n=144 mean±sd	95% CI[e]	P-value[f]
Height (cm)	128.22±13.20	135.66±12.67	-13.88,-0.99	0.02	129.54±7.66	135.86±11.08	-12.31,-0.33	0.04
Weight (kg)	25.55±6.15	31.72±10.00	-11.07,-1.26	0.01	26.46±3.77	31.66±8.45	-9.72,-0.68	0.02
Muscle mass[a] (kg)	4.71±2.99	8.74±4.98	-6.47,-1.59	0.001	5.74±3.60	9.88±4.33	-6.51,-1.78	0.001
Mid-arm muscle circumference[b] (mm)	17.95±1.72	20.27±2.70	-3.66,-0.98	0.001	17.92±1.14	20.31±2.68	-3.82,-0.96	0.001
Arm muscle area[b] (mm^2)	1818.66±320.90	2246.33±55.30	-698.25,-157.08	0.002	1869.36±268.28	2425.12±556.60	-853.59,-257.94	0.000
Adipose mass[a] (kg)	8.01±2.79	11.06±5.17	-5.58,-0.53	0.02	7.47±1.34	9.94±5.24	-5.25,0.31	0.08
% fat: tri/calf[c]	16.87±3.74	19.77±5.19	-5.47,-0.34	0.03	13.57±2.28	16.07±6.53	-5.98,0.98	0.16
% fat: tri/sub[c]	20.42±4.25	24.06±5.15	-6.21,-1.08	0.006	17.83±2.38	20.40±6.76	-6.17,1.02	0.16
Sum of 6 skinfolds[d] (mm)	54.62±16.53	70.83±29.45	-30.60,-1.81	0.03	48.86±7.58	60.77±33.57	-29.73,5.9	0.19

a (140)
b (141)
c percent body fat (142)
d triceps, subscapular, supraspinale, abdominal, thigh, calf
e confidence interval for the difference between the subjects and the reference group
f independent t-test of the difference between the subjects and the reference group

Cross-sectional

Skinfold and girth measurements

The measures of muscularity (MM, MAMC, AMA) and adiposity (AM, %fat, Σ6SF) of the males and females with CF aged 6 to 11 years were lower than the COGRO reference group immediately prior to the intervention (Table 5.15). Table 5.15 shows that the differences between the study and reference group were statistically significant except for the measures of adiposity of the males.

The z-scores of the anthropometric measures (Table 5.16) highlighted the magnitude of the deficits in muscularity and adiposity of the children with CF and the differences between genders. Table 5.16 shows that the deficit in the mean weight of the females with CF (z-score -0.91) was contributed to by slightly larger deficits in muscularity (MM, MAMC and AMA z-scores were -0.98 to -0.92), compared with adiposity (AM, %fat and Σ6SF z-scores were -0.85 to -0.58). The deficit in weight of the males with CF (z-score -0.53) was due to much greater deficits in muscularity (MM, MAMC and AMA z-scores were -1.04 to -0.93), compared with adiposity (AM, %fat and Σ6SF z-scores were -0.43 to -0.34). The differences between the z-scores for males and females with CF were statistically significant only for AM (p=0.01). The relationships between the anthropometric measurements and lung function were not statistically significant.

Table 5.16

Anthropometry of 6 to 11 year old pre-pubertal children with cystic fibrosis, standardised for age and gender against the COGRO reference group (143), immediately prior to the intervention

Measure	Females n=17 mean±sd	Males n=14 mean±sd	Estimate (95% CI)[e]	P-value[f]
z-height	-1.05±0.90	-0.52±0.81	-0.53 (-1.17, 0.10)	0.10
z-weight	-0.91±0.40	-0.53±0.65	-0.38 (-0.77, 0.01)	0.055
z-muscle mass[a]	-0.92±0.70	-1.03±0.97	0.11 (-0.51, 0.72)	0.72
z-mid-arm muscle circumference[b]	-0.98±0.52	-0.93±0.55	-0.05 (-0.44, 0.34)	0.79
z-arm muscle area[b]	-0.92±0.66	-1.04±0.62	0.12 (-0.36, 0.59)	0.62
z-adipose mass[a]	-0.85±0.45	-0.41±0.47	-0.44 (-0.77, -0.10)	0.013
z-% fat: triceps & calf[c]	-0.58±0.69	-0.43±0.61	-0.15 (-0.63, 0.34)	0.54
z-% fat: triceps & subscapular[c]	-0.73±0.78	-0.38±0.54	-0.35 (-0.85, 0.15)	0.17
z-sum of 6 skinfolds[d]	-0.58±0.58	-0.34±0.43	-0.24 (-0.62, 0.14)	0.20

a (140)

b (141)

c (142)

[d] triceps, subscapular, supraspinale, abdominal, thigh, calf
[e] estimate (confidence interval) for the difference between females and males
[f] independent t-test for the difference between females and males

Longitudinal

Height and weight

Statistically significant time trends only were observed for the raw measurements of weight, height and %IBW of the 2 to 11 year old children with CF (intervention and control group combined) (Table 5.17). Mean weight and height improved consecutively from baseline to 12 months follow-up, whereas mean %IBW varied over time. These time trends for weight and height were not significant after adjusting for covariates, but the time trend for %IBW remained significant.

Initially, there were no significant effects of time, group or group by time on z-height and z-weight measurements that had been standardised against the COGRO reference group (Table 5.17). However, after adjusting for the covariates, significant time trends were observed for both measurements (p=0.043 and p=0.028, respectively). Mean z-height and z-weight both varied slightly between baseline and the 6 month follow-up time point and then improved to a greater magnitude at 12 month follow-up. After adjusting for the covariates, a significant group by time effect was observed for z-height (p=0.022), but not for z-weight. The mean z-height values for both groups either remained the same or varied slightly by similar magnitudes over baseline to the 6 month follow-up time point, but from 6 to 12 months follow-up there was a larger improvement in the intervention group (by 0.26 standard deviations), whereas the mean value for the control group was slightly worse (by 0.02 standard deviations).

Longitudinal

Skinfolds and girths

Statistically significant time trends were observed for a majority of the raw anthropometric measures, skinfolds and girths (Tables 5.18 to 5.20), but only for the age- and gender-standardised measures of z-%adipose mass, z-supraspinale skinfold and z-thigh skinfold of the 5 to 11 year old children with CF (Tables 5.21 to 5.22). The statistically significant time trends for the three age- and gender-standardised measurements indicate that the children with CF were performing differently to the

reference group over time. The direction of the changes in the mean values of the measures varied considerably over time.

These time trends and the effects of group and group by time were all non-significant after adjusting for covariates. However, adjusting for covariates yielded other significant effects of time, namely for z-%fat (triceps,calf), z-calf skinfold and z-forearm girth (Tables 5.21 to 5.23). Mean values for z-%fat (triceps,calf), z-calf skinfold worsened over time, whereas mean z-forearm girth values improved over time.

Longitudinal

Lung function

The effects of group, time and group by time were all non-significant for the %predicted measures of lung function. The results are summarised in Table 5.24.

Table 5.17

Multivariate analysis of height and weight measurements of 2 to 11 year old children with cystic fibrosis in the intervention group (n=28) and control group (n=28)

Measure		F-ratio (d.f.)[g]	P-value
raw weight	Time[d]	106.88 (4, 51)	0.000
	Group[e]	0.60 (1, 54)	0.37
	Group*Time[f]	0.28 (4, 51)	0.80
raw height	Time	272.30 (4, 51)	0.000
	Group	0.83 (1, 53)	0.44
	Group*Time	0.42 (4, 51)	0.89
%IBW[a]	Time	3.62 (4, 51)	0.011
	Group	2.07 (1, 54)	0.16
	Group*Time	0.56 (4, 51)	0.69
z-weight[b]	Time	1.81 (4, 36)	0.15***
	Group	0.10 (1, 39)	0.75
	Group*Time	0.84 (4, 36)	0.51
z-height[b]	Time	1.27 (4, 36)	0.30***
	Group	0.16 (1, 39)	0.69
	Group*Time	1.00 (4, 36)	0.42***
z-weight[c]	Time	2.34 (4, 51)	0.07
	Group	0.08 (1, 54)	0.78
	Group*Time	0.24 (4, 51)	0.91
z-height[c]	Time	0.50 (4, 51)	0.74
	Group	0.78 (1, 54)	0.38
	Group*Time	0.64 (4, 51)	0.64

[a] percent ideal body weight (18)
[b] z-score standardised for age and gender against the COGRO reference group (143)
[c] z-score standardised for age and gender against the NCHS reference group(136)
[d] main effect of time on the measure (intervention and control group combined)
$H_0: \mu_1=\mu_2=\mu_3=\mu_4=\mu_5$
Wilks' Lambda multivariate test
[e] main effect of group over all five time points, ie average group effect
H_0: average difference between groups over all five time points = 0
Repeated measures analysis of variance
[f] difference of the measure between groups over all five time points
$H_0: \Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$
Wilks' Lambda multivariate test
[g] numerator degrees of freedom, denominator/error degrees of freedom
*** $p < 0.05$ after adjusting for the covariates

Table 5.18

Multivariate analysis of raw anthropometric measures of 5 to 11 year
old children with cystic fibrosis in the intervention group (n=20) and
control group (n=18)

Raw anthropometric measure		F-ratio (d.f.)[j]	P-value
muscle mass[a]	Time[g]	30.13 (4, 32)	0.000
	Group[h]	0.05 (1, 35)	0.83
	Group*Time[i]	0.71 (4, 32)	0.59
mid-arm muscle circumference[b]	Time	27.99 (4, 33)	0.000
	Group	0.20 (1, 36)	0.66
	Group*Time	0.54 (4, 33)	0.71
arm muscle area[b]	Time	26.31 (4, 33)	0.000
	Group	0.10 (1, 36)	0.76
	Group*Time	0.20 (4, 33)	0.93
adipose mass[a]	Time	24.5 (4, 33)	0.000
	Group	0.01 (1, 36)	0.98
	Group*Time	0.56 (4, 33)	0.69
% fat: tricep & calf[c]	Time	1.23 (4, 33)	0.32
	Group	0.60 (1, 36)	0.44
	Group*Time	0.29 (4, 33)	0.88
% fat: tricep & subscapular[c]	Time	3.01 (4, 33)	0.032
	Group	0.87 (1, 36)	0.36
	Group*Time	0.64 (4, 33)	0.63
% adipose mass	Time	9.35 (4, 33)	0.000
	Group	0.13 (1, 36)	0.72
	Group*Time	0.95 (4, 33)	0.45
sum 4 skinfolds[d]	Time	2.40 (4, 33)	0.07
	Group	0.32 (1, 36)	0.58
	Group*Time	0.82 (4, 33)	0.52

sum 6 skinfolds[e]	Time	3.20 (4, 33)	0.025
	Group	0.26 (1, 36)	0.61
	Group*Time	0.51 (4, 33)	0.73
sum 8 skinfolds[f]	Time	4.32 (4, 33)	0.006
	Group	0.23 (1, 36)	0.63
	Group*Time	0.51 (4, 33)	0.73
sum 4 height-corrected skinfolds[d]	Time	2.62 (4, 33)	0.052
	Group	0.39 (1, 36)	0.54
	Group*Time	0.97 (4, 33)	0.44
sum 6 height-corrected skinfolds[e]	Time	2.88 (4, 33)	0.038
	Group	0.33 (1, 36)	0.57
	Group*Time	0.53 (4, 33)	0.71
sum 8 height-corrected skinfolds[f]	Time	2.78 (4, 33)	0.04
	Group	0.29 (1, 36)	0.59
	Group*Time	0.56 (4, 33)	0.70

a (140)

b (141)

c (142)

[d] triceps, subscapular, biceps, supraspinale

[e] triceps, subscapular, supraspinale, abdominal, thigh, calf

[f] triceps, subscapular, biceps, iliac crest, supraspinale, abdominal, thigh, calf

[g] main effect of time on the measure (intervention and control group combined)

H_0: $\mu_1 = \mu_2 = \mu_3 = \mu_4 = \mu_5$

Wilks' Lambda multivariate test

[h] main effect of group over all five time points, ie average group effect

H_0: average difference between groups over all five time points = 0

Repeated measures analysis of variance

[i] difference of the measure between groups over all five time points

H_0: $\Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$

Wilks' Lambda multivariate test

[j] numerator degrees of freedom, denominator/error degrees of freedom

Table 5.19

Multivariate analysis of raw skinfold measurements of 5 to 11 year old children
with cystic fibrosis in the intervention group (n=20) and control group (n=18)

Raw skinfold measurement		F-ratio (d.f.)[d]	P-value
tricep	Time[a]	3.84 (4, 33)	0.011
	Group[b]	0.19 (1, 36)	0.66
	Group*Time[c]	0.69 (4, 33)	0.60
subscapular	Time	0.85 (4, 33)	0.50
	Group	0.03 (1, 36)	0.87
	Group*Time	0.74 (4, 33)	0.57
bicep	Time	3.06 (4, 33)	0.03
	Group	0.39 (1, 36)	0.54
	Group*Time	0.58 (4, 33)	0.68
iliac	Time	11.56 (4, 33)	0.000
	Group	0.004 (1, 36)	0.95
	Group*Time	0.74 (4, 33)	0.57
supraspinale	Time	2.63 (4, 33)	0.052
	Group	0.89 (1, 36)	0.35
	Group*Time	0.47 (4, 33)	0.76
abdominal	Time	6.36 (4, 33)	0.001
	Group	1.10 (1, 36)	0.30
	Group*Time	0.30 (4, 33)	0.88
thigh	Time	6.58 (4, 33)	0.001
	Group	0.01 (1, 36)	0.97
	Group*Time	1.05 (4, 33)	0.40
calf	Time	0.33 (4, 33)	0.86
	Group	0.24 (1, 36)	0.62
	Group*Time	0.22 (4, 33)	0.92

[a] main effect of time on the measure (intervention and control group combined)

H_0: $\mu_1=\mu_2=\mu_3=\mu_4=\mu_5$

Wilks' Lambda multivariate test

[b] main effect of group over all five time points, ie average group effect

H_0: average difference between groups over all five time points = 0

Repeated measures analysis of variance

[c] difference of the measure between groups over all five time points

H_0: $\Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$

Wilks' Lambda multivariate test

[d] numerator degrees of freedom, denominator/error degrees of freedom

Table 5.20

Multivariate analysis of raw girth measurements of 5 to 11 year old children with cystic fibrosis in the intervention group (n=20) and control group (n=18)

Raw girth measurement		F-ratio (d.f.)[d]	P-value
relaxed arm	Time[a]	26.09 (4, 33)	0.000
	Group[b]	0.20 (1, 36)	0.66
	Group*Time[c]	0.57 (4, 33)	0.69
flexed arm	Time	29.58 (4, 33)	0.000
	Group	0.15 (1, 36)	0.70
	Group*Time	0.87 (4, 33)	0.49
forearm	Time	42.45 (4, 33)	0.000
	Group	0.08 (1, 36)	0.78
	Group*Time	1.40 (4, 33)	0.25
wrist	Time	27.88 (4, 33)	0.000
	Group	0.11 (1, 36)	0.74
	Group*Time	0.46 (4, 33)	0.76
chest	Time	184.91 (4, 33)	0.000
	Group	0.01 (1, 36)	0.95
	Group*Time	0.81 (4, 33)	0.53
waist	Time	23.66 (4, 33)	0.000
	Group	0.01 (1, 36)	0.98
	Group*Time	0.54 (4, 33)	0.71
hip	Time	76.58 (4, 33)	0.000
	Group	0.12 (1, 36)	0.73
	Group*Time	0.97 (4, 33)	0.90
thigh	Time	64.41 (4, 32)	0.000
	Group	0.01 (1, 35)	0.95
	Group*Time	0.44 (4, 32)	0.78

calf	Time	69.60 (4, 33)	0.000
	Group	0.05 (1, 36)	0.82
	Group*Time	0.32 (4, 33)	0.86
ankle	Time	53.00 (4, 33)	0.000
	Group	0.41 (1, 36)	0.52
	Group*Time	0.38 (4, 33)	0.82

[a] main effect of time on the measure (intervention and control group combined)

H_0: $\mu_1 = \mu_2 = \mu_3 = \mu_4 = \mu_5$

Wilks' Lambda multivariate test

[b] main effect of group over all five time points, ie average group effect

H_0: average difference between groups over all five time points = 0

Repeated measures analysis of variance

[c] difference of the measure between groups over all five time points

H_0: $\Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$

Wilks' Lambda multivariate test

[d] numerator degrees of freedom, denominator/error degrees of freedom

Table 5.21

Multivariate analysis of anthropometric measures of 5 to 11 year old children with cystic fibrosis in the intervention group (n=20) and control group (n=18), standardised against the GOGRO reference group (143)

Z- anthropometric measure		F-ratio (d.f.)[h]	P-value
z-muscle mass[a]	Time[e]	2.54 (4, 32)	0.06
	Group[f]	0.03 (1, 35)	0.86
	Group*Time[g]	0.61 (4, 32)	0.66
z-mid-arm muscle circumference[b]	Time	0.76 (4, 33)	0.56
	Group	2.24 (1, 36)	0.14
	Group*Time	1.14 (4, 33)	0.36
z-arm muscle area[b]	Time	0.71 (4, 33)	0.59
	Group	1.17 (1, 36)	0.29
	Group*Time	0.50 (4, 33)	0.74
z-adipose mass[a]	Time	1.48 (4, 33)	0.23
	Group	0.96 (1, 36)	0.33
	Group*Time	2.00 (4, 33)	0.12
z-% fat: tricep & calf[c]	Time	0.61 (4, 33)	0.66***
	Group	0.48 (1, 36)	0.49
	Group*Time	0.68 (4, 33)	0.61
z-% fat: tricep & subscapular[c]	Time	0.93 (4, 33)	0.46
	Group	1.12 (1, 36)	0.30
	Group*Time	1.60 (4, 33)	0.20
z-% adipose mass	Time	3.44 (4, 33)	0.019
	Group	0.02 (1, 36)	0.88
	Group*Time	1.93 (4, 33)	0.13

(continued next page)

z-sum 6 skinfolds[d]	Time	1.94 (4, 33)	0.13
	Group	0.14 (1, 36)	0.71
	Group*Time	1.24 (4, 33)	0.31
z-sum 6 height-corrected skinfolds[d]	Time	1.90 (4, 33)	0.13
	Group	0.02 (1, 36)	0.88
	Group*Time	1.08 (4, 33)	0.38

a (140)

b (141)

c (142)

[d] triceps, subscapular, supraspinale, abdominal, thigh, calf

[e] main effect of time on the measure (intervention and control group combined)

    H_0: $\mu_1 = \mu_2 = \mu_3 = \mu_4 = \mu_5$

    Wilks' Lambda multivariate test

[f] main effect of group over all five time points, ie average group effect

    H_0: average difference between groups over all five time points = 0

    Repeated measures analysis of variance

[g] difference of the measure between groups over all five time points

    H_0: $\Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$

    Wilks' Lambda multivariate test

[h] numerator degrees of freedom, denominator/error degrees of freedom

*** $p < 0.05$ after adjusting for the covariates

Table 5.22

Multivariate analysis of skinfold measurements of 5 to 11 year old children with cystic fibrosis in the intervention group (n=20) and control group (n=18), standardised against the GOGRO reference group (143)

Z-skinfold measurement		F-ratio (d.f.)[d]	P-value
z-tricep	Time[a]	1.31 (4, 33)	0.29
	Group[b]	0.66 (1, 36)	0.42
	Group*Time[c]	0.70 (4, 33)	0.60
z-subscapular	Time	1.86 (4, 33)	0.14
	Group	0.29 (1, 36)	0.59
	Group*Time	0.81 (4, 33)	0.53
z-supraspinale	Time	4.29 (4, 33)	0.007
	Group	0.30 (1, 36)	0.59
	Group*Time	1.55 (4, 33)	0.21
z-abdominal	Time	0.72 (4, 33)	0.58
	Group	0.01 (1, 36)	0.94
	Group*Time	1.03 (4, 33)	0.41
z-thigh	Time	4.21 (4, 33)	0.007
	Group	0.01 (1, 36)	0.95
	Group*Time	1.41 (4, 33)	0.25
z-calf	Time	1.32 (4, 33)	0.28***
	Group	0.10 (1, 36)	0.75
	Group*Time	0.58 (4, 33)	0.68

[a] main effect of time on the measure (intervention and control group combined)

H_0: $\mu_1=\mu_2=\mu_3=\mu_4=\mu_5$

Wilks' Lambda multivariate test

[b] main effect of group over all five time points, ie average group effect

H_0: average difference between groups over all five time points = 0

Repeated measures analysis of variance

[c] difference of the measure between groups over all five time points

H_0: $\Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$

Wilks' Lambda multivariate test

[d] numerator degrees of freedom, denominator/error degrees of freedom

*** $p<0.05$ after adjusting for the covariates

Table 5.23

Multivariate analysis of girth measurements of 5 to 11 year old children with cystic fibrosis in the intervention group (n=20) and control group (n=18), standardised against the GOGRO reference group (143)

Z- girth measurement		F-ratio (d.f.)[d]	P-value
z-relaxed arm	Time[a]	0.81 (4, 33)	0.53
	Group[b]	2.57 (1, 36)	0.12
	Group*Time[c]	1.34 (4, 33)	0.28
z-forearm	Time	2.21 (4, 33)	0.09***
	Group	1.20 (1, 36)	0.28
	Group*Time	1.37 (4, 33)	0.27
z-chest	Time	1.42 (4, 33)	0.25
	Group	0.17 (1, 36)	0.68
	Group*Time	1.33 (4, 33)	0.28
z-thigh	Time	1.60 (4, 32)	0.20
	Group	0.00 (1, 35)	0.99
	Group*Time	1.37 (4, 32)	0.26
z-calf	Time	2.65 (4, 33)	0.051
	Group	0.21 (1, 36)	0.65
	Group*Time	0.94 (4, 33)	0.46

[a] main effect of time on the measure (intervention and control group combined)
    H_0: $\mu_1 = \mu_2 = \mu_3 = \mu_4 = \mu_5$
    Wilks' Lambda multivariate test
[b] main effect of group over all five time points, ie average group effect
    H_0: average difference between groups over all five time points = 0
    Repeated measures analysis of variance
[c] difference of the measure between groups over all five time points
    H_0: $\Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$
    Wilks' Lambda multivariate test
[d] numerator degrees of freedom, denominator/error degrees of freedom
*** p<0.05 after adjusting for the covariates

Table 5.24

Multivariate analysis of %predicted lung function measurements (148, 149) of 6 to 11 year old children with cystic fibrosis in the intervention group (n=16) and control group (n=16)

Measure		F-ratio (d.f.)[g]	P-value
FEV_1[a]	Time[d]	1.85 (4, 27)	0.15
	Group[e]	0.27 (1, 30)	0.61
	Group*Time[f]	0.36 (4, 27)	0.84
$FEF_{25\ to\ 75\%}$[b]	Time	0.92 (4, 27)	0.47
	Group	0.44 (1, 30)	0.51
	Group*Time	0.20 (4, 27)	0.94
FVC[c]	Time	1.42 (4, 27)	0.25
	Group	0.27 (1, 30)	0.61
	Group*Time	0.52 (4, 27)	0.72

[a] forced expiratory volume in 1 second
[b] forced expiratory flow at 25% to 75 % of vital capacity
[c] forced vital capacity
[d] main effect of time on the measure (intervention and control group combined)
    H_0: $\mu_1=\mu_2=\mu_3=\mu_4=\mu_5$
    Wilks' Lambda multivariate test
[e] main effect of group over all five time points, ie average group effect
    H_0: average difference between groups over all five time points = 0
    Repeated measures analysis of variance
[f] difference of the measure between groups over all five time points
    H_0: $\Delta T_1 = \Delta T_2 = \Delta T_3 = \Delta T_4 = \Delta T_5$
    Wilks' Lambda multivariate test
[g] numerator degrees of freedom, denominator/error degrees of freedom

Discussion

This is one of the first reports to describe extensive anthropometric data obtained from children with CF who have mild to moderate lung disease. The subjects were without secondary conditions known to adversely affect nutritional status (short-gut syndrome and liver disease) and the mean %IBW values for both genders were within the normal range for CF (90 to 110% IBW) (18). Our finding of a large deficit in the muscularity of the children with CF, indicative of malnutrition, suggests that using only height, weight and the %IBW index to assess nutritional status could be misleading.

Of particular interest, the cross-sectional analysis revealed that the deficit in the muscularity (MM, MAMC, AMA) of the children with CF was much greater than the deficit in adiposity (AM, %fat and Σ6SF) (Tables 5.15 and 5.16). This finding was surprising as it is opposite to that observed in uncomplicated protein-energy malnutrition, where the adaptive response to under-nutrition in those without CF usually results in protein being spared at the expense of fat (159). If the children with CF in our study were exhibiting straightforward malnutrition then it would have been reasonable to expect the deficit in muscularity to be less than, or at most similar to, the deficit in adiposity.

As expected, statistically significant time trends were observed for most of the raw measurements of height, weight, skinfolds, girths and anthropometric measures in the longitudinal analysis of the data (Tables 5.17 to 5.20). The magnitude of the effect of these time trends was reduced when the measurements were standardised for age and gender against the COGRO reference group. There were no statistically significant group or group by time effects in the age- and gender-standardised measurements without adjustment for covariates.

After adjusting for covariates, the time trends were non-significant for all the standardised measurements except for z-%fat (triceps,calf), z-calf skinfold and z-forearm girth. Adjusting for covariates revealed a significant effect of the *Go and Grow with CF* program on only one anthropometric measurement, namely height

standardised against the COGRO reference group (Group by Time p=0.022). From the 6 to 12 month follow-up time points, the mean z-height value of the intervention group improved and the value of the control group was worse.

The significant effects of time for the raw, and raw height-corrected, sum of 6 and 8 skinfolds and not 4 skinfolds (Table 5.18) potentially supports the recommendation of anthropometrists to measure a greater range of anatomical sites in order for measures to be more representative of actual body composition and to detect changes. However, comparison with a superior method (eg. dual-energy x-ray absorptiometry) is required before any conclusions can be drawn.

The deficit in the muscularity of the children in the clinical trial may be the consequence of individuals with CF being in a state of chronic catabolic stress and starvation (36, 51, 159, 160). This chronic catabolic state may be due to a CF-related increase in muscle protein catabolism (159, 160), decrease in protein synthesis (160), alteration in digestion and/or the additional stress of chronic pulmonary disease and infections (36) which may cause an increase in energy expenditure and reduce appetite. It is possible that any, or a combination, of these factors could be responsible for both retarding growth and causing detrimental effects on the respiratory diaphragm, the accessory inspiratory and expiratory muscles and the lung tissue growth of children with CF (23, 77, 161). In a review of the role of nutrition and exercise training in CF, Heijerman (162) suggests that nutrition interventions may be most effective when administered in early childhood when the airways are still capable of growth.

A strong relationship between maintaining good nutritional status throughout childhood and the preservation of pulmonary function in CF has not yet been described. This may be because most studies have relied on height and weight only as indicators of nutritional status (13, 16, 17). The small sample size, large inter-subject variability, relatively narrow age range and reasonably well-maintained respiratory function of the subjects may have limited the ability of the present study to indicate any association between the anthropometric measures and lung function.

Other studies have found metabolic measures of body composition to be good predictors of pulmonary function in CF (23). But, as with many of the methods used to assess body composition, total body potassium analysis is not widely available and is not suitable for regular monitoring of children who have CF. Anthropometry appears to be the best option for routine use in a CF clinic as it is non-invasive, inexpensive, portable and the measurements can be obtained quickly by a trained anthropometrist (it takes about 15 minutes to measure in triplicate the 8 skinfolds and 10 girths used in this study). Variations in the skin compressibility, skin thickness and tissue patterning of individuals can be accounted for by measuring numerous sites on the upper and lower body, trunk and limbs (55-57). The reliability and validity of anthropometry can be further enhanced by adhering to standard procedures when taking the measurements (135).

The longitudinal study of the body composition of children with CF by Stettler et al (2) illustrates the limitations of relying on one site to indicate changes in fat free mass. The mean upper-arm muscle area of the children with CF (as determined by anthropometry) was not statistically significantly different to that of the control group, but fat-free mass of the boys with CF (as determined by skinfolds from four sites, total body water and total body electrical conductivity) were lower (2).

5.7 Summary

The main findings from the clinical trial are listed below.

Process evaluation

- ◆ There was a high level of participant satisfaction, perceived learning and recommendation of the program to other families who have a child with CF.
- ◆ The factors which may have contributed to carers' motivation to complete the home-based course include a desire to learn, flexibility of the program, monthly telephone contact with the dietitian and fortnightly newsletters.

Knowledge and self-management

♦ The level of knowledge of children and carers about fat, energy and salt was poor.

♦ *Go and Grow with CF* was effective in increasing children's knowledge in the short-term.

♦ It may be necessary to provide periodic nutrition and PERT education to assist the retention of knowledge gains from the program.

♦ Both appropriate and inappropriate strategies are commonly used to self-manage malabsorption, food and enzyme refusal and teasing about pancreatic enzymes.

Behaviour

♦ Use of the BPFAS revealed a statistically significant higher rate of inappropriate child meal-time behaviours in younger children with CF.

♦ The moderate frequency of adverse food-related behaviours of children with CF supports the need for behaviour management programs.

♦ Immediately post-intervention, fewer carers in the intervention group endorsed items in the carers' response section as problematic.

Dietary intake

♦ The dietary intake of the children with CF in the clinical trial was possibly grossly overestimated using the GPHNS FFQ.

Body composition, using anthropometry

♦ The children with CF were found to have a large deficit in muscularity, indicative of malnutrition.

♦ The deficit in muscularity was much greater than the deficit in adiposity.

♦ Use of height, weight and the %IBW index to assess nutritional status could be misleading.

♦ Extensive anthropometry appears to be the best option for routine use in a CF clinic.

6. CHAPTER SIX: CLINICAL IMPLEMENTATION

The study described in this section evolved after

- the process evaluation of the pilot *Go and Grow with CF* program indicated the suitability of the intervention for clinic-wide implementation, and

- inclusion of the Australian PERT Guidelines in the revised version of the program provided an opportunity to assess fat-based dosing.

The *Go and Grow with CF* program was revised and updated in 1998, based on the process evaluation of the pilot program (Section 5.2) and recent advances in CF nutrition and PERT recommendations. The weekly activities of the revised home-based course are listed in Appendix 3.1. Modifications to the *Go and Grow with CF* home-based course included the following:

1. More comprehensive information about using a reward system as process evaluation of the pilot program indicated that carers were not consistently using rewards and did not understand their importance for achieving behavioural change in children.

2. Guidelines for using *Go and Grow with CF* with children who have pancreatic sufficiency as some topics in the program are relevant (energy and salt) and others are not (malabsorption and PERT).

3. Suggestions for involving other family members as some carers who participated in the pilot program voluntarily indicated that *Go and Grow with CF* was useful for educating siblings about the therapeutic dietary needs of the child with CF, such that they were less opposed to apparent meal and snack inequalities.

4. The addition of behavioural learning activities regarding the Australian PERT Guidelines (67, 68); and a dietary fat target for primary school-aged children with CF of at least 100 g per day (12), which were published after the pilot program had been implemented (Appendix 3.1).

The Australian PERT Guidelines for CF differ from those of other countries in that the dosage recommended for each meal and snack is dependent on the amount of dietary fat consumed, rather than on body weight. These fat-based doses are expressed as international units (IU) of lipase per gram of dietary fat, and more simply for families as one capsule per a specific number of grams of fat, depending on the type of pancreatic enzyme preparation used and the individual's need (67, 68). For example, a dose of 1 000 IU of lipase per gram of fat is expressed as one capsule per 5 g of fat for preparations containing 5 000 IU of lipase per capsule, and as one capsule per 10 g of fat for preparations containing 10 000 IU of lipase per capsule. Although research assessing the different methods of pancreatic enzyme dosing is scarce (39, 163, 164), the fat-based method is physiologically sound (69), and preferred (39), and warrants further investigation. Clinical implementation of the revised *Go and Grow with CF* program provided an opportunity to investigate the effects of fat-based PERT dosing on fat absorption in children with CF, while conducting further process evaluation.

Methods

Participants

All children aged 2 to 13 years attending the CF clinic at Princess Margaret Hospital (n=97) and/or their parents were given the opportunity to participate in the *Go and Grow with CF* program between April and July 1998, in order to learn the Australian PERT Guidelines. Even though *Go and Grow with CF* targets 2 to 11 year olds and their parents, children aged 12 and 13 years of age were also offered the 1998 revised version as the majority of these adolescents (10 out of 12) had participated in the clinical trial which evaluated the original program. It was considered that children who were in the intervention group during the clinical trial would benefit from participation in the revised program due to factors related to improvements and additions to *Go and Grow with CF*, as listed in the introduction to Chapter 6.

Information about how the change in PERT practices at Princess Margaret Hospital would be assessed was provided by post to children and their parents at the same time that they received an invitation to participate in the home-based *Go and Grow*

with CF program. The information letter asked parents to consent to an assessment of their child's level of FFE both pre- and post-participation in the *Go and Grow with CF* program. The parent/s who accompanied their child to the next scheduled CF clinic appointment was then approached by the investigator to determine the family's level of involvement in *Go and Grow with CF* and the assessment of fat absorption.

Process evaluation was conducted with children and their carers in the *Go and Grow with CF* target group age range only (ie. 2 to 11 year olds). As there were no exclusion criteria, all 77 children with CF aged 2 to 11 years and/or their carers were eligible to participate in the clinic-wide implementation of the revised *Go and Grow with CF* program. Potential program participants included 58 children aged 6 to 11 years and 73 carers of children aged 2 to 11 years.

Parents of a self-selected cohort of 38 children aged between 2 and 13 years, who were assessed as having pancreatic insufficiency when CF was diagnosed, agreed to a range of measurements being taken (eg. height, weight, dietary and pancreatic enzyme intake and faecal output) approximately six months apart, pre- and post-implementation of the Australian PERT Guidelines. Each child acted as his/her own control. Comparisons were made between pre- and post-intervention intakes of fat and lipase, fat absorption, height and weight.

6.1 Process evaluation

Materials

The clinic-wide implementation of the revised *Go and Grow with CF* program is outlined in Appendix 6.1.

Participation rates were calculated as a proportion of the potential program participants. Carers' program goals for themselves, and for their children, were

documented during the introductory workshops, at CF clinic appointments or posted to the dietitian (me) from home.

A dietitian with CF expertise (MP), but not the program developer, collected all subsequent information from carers for process evaluation of the revised program (Appendix 6.2). During telephone contact with the dietitian (MP) at weeks 4, 8 and 12 of the home-based course, carers self-reported what they, and what they thought their child, were learning, enjoying and disliking by answering open-ended questions. On each occasion carers were asked for suggestions to improve the program. At week 4, carers were also asked to indicate their level of interest in *Go and Grow with CF* prior to commencing the program and their level of motivation to keep doing the program. At weeks 4 and 8, carers were asked if they were using the reward system with their child. At week 12, carers were asked to indicate the topics or activities in the program that were most beneficial, their preference for calculating PERT dosages, what helped them to continue participating in the home-based course, and whether they would recommend *Go and Grow with CF* to other families who have a child with CF. At the completion of *Go and Grow with CF*, carers indicated if the program goals they set at the beginning of the program were achieved. Telephone contact was made one month after the concluding workshop with carers who had not completed the home-based course by week 12 of the program. The amount of dietetic time spent conducting all aspects of *Go and Grow with CF* was recorded. Use of the *Go and Grow with CF* program by non-participants after the clinic-wide implementation was recorded.

Results

Participation

Workshop and home-based course participation rates for the revised program are summarised in Tables 6.1 and 6.2, including comparisons with the pilot program.

Table 6.1

Participation rates for the *Go and Grow with CF* workshops

| | Revised program | | Pilot program |
	Children aged 6 to 11 years n=58	Carers of children aged 2 to 11 years n=73	Carers 1996 n=27
Introductory workshop	29%	48%	60%
Concluding workshop	10%	11%	60%

Table 6.2

Participation rates for the *Go and Grow with CF* home-based course

| | Revised program | | Pilot program |
	Children aged 6 to 11 years n=58	Carers of children aged 2 to 11 years n=73	Carers 1996 n=27
Completed on schedule	27%	30%	72%
Completed 1 month later	21%	23%	10%
Part-completed	26%	21%	10%
Did not participate	26%	26%	8%

Only 29% of children aged 6 to 11 years and 48% of carers of 2 to 11 year olds attended the introductory workshop (Table 6.1). Over half the 6 to 11 year old children who attended the workshops were accompanied by at least one sibling.

Nineteen carers (26%) indicated they would not be participating in the home-based course (Table 6.2). The reason given for non-participation by over two-thirds of these carers was that they were too busy. Other carers chose not to participate in *Go and Grow with CF* because they perceived their child did not have any nutrition or PERT problems, the family was moving interstate or going on an extended holiday. Of the carers who indicated they would not be participating in the program (n=19), the majority (n=15, 79%) had a child with CF in the age range for which *Go and Grow with CF* was especially written (ie. 6 to 11 year olds), nearly half (n=8, 42%) had children who were nutritionally at risk (based on inadequate weight and height gains over the previous year and who had not responded to non-invasive nutrition interventions), one-quarter (n=5) had children who irregularly attend the CF clinic (<twice/year), and only one-fifth (n=4) had completed the pilot *Go and Grow with CF* program. In contrast, only 10% (n=4) of children whose carers completed the home-based course had experienced inadequate weight and height gains over the previous year.

Twenty-seven percent of school-aged children and 30% of carers completed the home-based course on schedule, ie by the end of the concluding workshop (Table 6.2). Other commitments temporarily interrupted some families. One month after the concluding workshop a total of 48% of children aged 6 to 11 years and 53% of carers had completed the home-based course (Table 6.2). Half the carers who participated in the pilot program also completed the revised program.

Twenty-six percent of school-aged children and 21% of carers started but did not complete the home-based course by one month after the concluding workshop (Table 6.2). Reasons volunteered by carers for non-completion were related to illness or being too busy. Most of these carers expressed their intention to continue

doing the activities in order to eventually complete the program. Nearly one-third of school-aged children and carers who started but did not complete the home-based course had participated in the pilot *Go and Grow with CF* program.

Only 10% of children aged 6 to 11 years and 11% of carers attended the concluding workshop (Table 6.1).

Four patients who were eligible to participate in *Go and Grow with CF* had pancreatic sufficiency. Carers of half these children indicated their non-intention to participate in the program, whereas the other half completed the home-based course.

Carers of all children diagnosed with CF over the previous four years attended the introductory workshop and 81% of these carers completed the home-based course.

Process evaluation

The process evaluation results which follow are for the group of 74% of carers of 2 to 11 year olds and their children who did some or all of the home-based course, unless otherwise stated. Most results were similar to the evaluation of the pilot *Go and Grow with CF* program. Only the findings that differed from the previous assessment of the program or were significant are reported.

Learning: Nutrition and enzyme concepts that carers reported they and their children learned were similar to the findings from the pilot study with particular emphasis on the additions to the revised program (matching enzymes to fat in food and drinks, achieving a fat target of 100 g or more per day by primary school-aged children and discovering the salt content of food and drinks).

Beneficial: Carers who completed the home-based course indicated that matching enzymes to fat in food and drinks was the most beneficial activity for them (63%) and their children (36%). The next highest rating for being most beneficial was the energy and fat topic (carers 26% and children 19%).

Dislikes: Some carers reported that they and their children disliked spending time calculating enzyme doses based on the fat content of the meal or snack.

Enzyme dose preferences: Ninety-four percent of carers who completed the home-based course reported that they preferred the new method of calculating enzyme doses, based on the fat content of meals and snacks, over the previous method, which was based on body weight. Carers' reasons for choosing to continue matching enzymes to fat in food and drinks were related to the improved symptoms in their child, such as less abdominal pain, less oily or loose bowel actions and less constipation; increased confidence with enzyme therapy; and ability to decrease the dose at some meals and snacks, without adverse effects. The three carers (6%) who indicated a preference for the previous method of pancreatic enzyme dosing, because it was easier to remember, also volunteered their intention to become more familiar with the new method.

Reinforcement: Seventy-two percent of carers who completed the home-based course reported using the weekly reward system to reinforce children's participation in the program. Twenty percent of carers reported they did not need to use the reward system as their child appeared to enjoy completing the worksheets and/or spending time with their carer. The remaining 8% of carers stated the reward system did not work with their child, but could not specify why. Fortnightly newsletters, monthly telephone contact with the dietitian and conversations with CF clinic staff motivated carers (39%, 35% and 27%, respectively) to complete the home-based course.

Suggestions for improving the program: One-third of carers made suggestions for improving the program. A majority indicated that the time allowed for completion of the home-based course should be increased or unlimited. Other carers suggested expanding the explanation about using the program with preschool-aged children. At the beginning of the program, some carers recommended including snack and recipe ideas. Interestingly, this information was planned for the newsletters that families would receive later in the home-based course.

Recommendation to others: Similar to the evaluation of the pilot program, all carers who completed the home-based course indicated they would recommend *Go and Grow with CF* to other families with a child who has CF. Carers reasons for recommending *Go and Grow with CF* were that the program:

- provided detailed information and the rationale for nutrition and PERT requirements
- explained how to achieve the child's needs
- helped carers and children develop nutrition and PERT skills
- helped increase confidence and independence
- was aimed at children and was fun to do, and
- prompted families to talk about CF together.

Dietetic time: An average of two and a half hours per family was spent by the dietitian conducting the *Go and Grow with CF* program over four months.

Follow-up: Relevant sections of the *Go and Grow with CF* home-based course were used to address current individual nutritional problems with three children who did not participate in the clinic-wide implementation of the program.

Discussion

The process evaluation of the revised version of *Go and Grow with CF* indicated that carers were highly satisfied with the program. Carers' goals, perceptions about their and their child's learning, enjoyment, dislikes and recommendation to others about the program were similar to those obtained during the pilot study. Matching enzymes to fat in food and drinks and the fat target for primary school-aged children of at least 100 g per day featured highly as the new concepts that carers perceived that they and their children learned. Carers also rated these activities as the most beneficial in the program.

Although the participation rates for the revised program were lower than those for the pilot program, it is likely that nearly two-thirds of the target group at Princess

Margaret Hospital have had considerable exposure to the *Go and Grow with CF* program over the past two years as some of the non-participants had completed the pilot program. These carers may have felt that they had learned sufficient information and could not devote time to participate in the program again. Indeed, being too busy was the most common reason for non-participation, even though *Go and Grow with CF* had been introduced as a home-based course which could be completed as time permitted. Some families may not have participated because CF education was not a high priority for them. Some were families with children who attend CF clinics on an irregular basis (<twice/year) and who had not volunteered to participate in the earlier clinical trial evaluating a change in nutrition services at Princess Margaret Hospital. It is interesting to note that most of the non-participants would have benefited from the intervention program because of the children's poor nutritional status despite previous attempts to intervene and limited education in the past, due to irregular CF clinic attendance.

Half the carers who did participate in the pilot *Go and Grow with CF* program also completed the revised home-based course. These carers appeared to be striving to ensure that their child would achieve and maintain optimum nutritional status. They hoped their child would gain a little more knowledge, now that they were two years older, and wanted to keep up-to-date with nutrition and PERT recommendations.

Of further interest is the finding that carers of all children diagnosed with CF in the past four years attended the introductory workshop and 81% completed the home-based course. This suggests families are most receptive to, and motivated about CF education, within the first few years after diagnosis.

Some carers reported that one of the things they and their children disliked about the revised *Go and Grow with CF* program was the time taken to calculate pancreatic enzyme doses based on the fat content of meals and snacks. However, most carers who completed the home-based course reported they preferred this method of enzyme dosing over the previous method which was based on body weight, because of the improved health of their child, increased confidence and the occasional need

for less capsules. Even the few carers who said they preferred the previous method of enzyme dosing, because it was easier to remember, also stated their intention to become more familiar with the new method.

Carers who completed the home-based course indicated that the fortnightly newsletters motivated them the most to keep participating in the revised program, whereas monthly telephone calls were rated the highest during the pilot study. This change in rating is likely to have been because the revised program newsletters contained numerous meal and snack ideas, which carers had indicated they were seeking in a nutrition program for children with CF. In addition, carers' reduced preference for telephone contact during the revised program may have been because the call at week 4 was relatively long. Faecal fat balance studies had been conducted with half the children involved in the process evaluation in order to assess the effect of the new Australian PERT Guidelines on absorption (Section 6.2). Consequently, telephone contact at week 4 for many carers included the provision of individualised dietary and PERT guidance based on laboratory test results, in addition to the collection of data for the process evaluation.

At the end of the revised program, carers indicated that conversations with CF clinic staff, such as doctors, nurses and the physiotherapist, had been a major motivational factor for continued participation. This higher rating for reinforcement from health-care providers in the revised program compared to the pilot study may have been because staff were now more familiar with the program and felt comfortable initiating conversation with families when prompted by a sheet about *Go and Grow with CF* that was attached to the medical records of the participating children.

A few carers suggested that the program could be improved by increasing the explanation about implementing *Go and Grow with CF* with pre-school-aged children. It was intended that carers of preschool-aged children would gain confidence in meeting the nutritional and PERT needs of their child with CF by completing the weekly activities in the home-based course themselves and thereby learn how to achieve optimum nutrition and PERT through each stage of their child's

development. Although written and verbal explanations for using the home-based course with children of various age categories were provided, some carers of young children decided to read the information only. Non-completion of the weekly activities may have limited carers' understanding and/or practice of the concepts in the program. During future implementations of *Go and Grow with CF* it may be beneficial to spend extra time with carers of preschoolers, in the introductory workshop or by telephone, exploring in detail how to maximise the use of the home-based course.

6.2 Fat-based pancreatic enzyme dosing

Measurements and materials

Faecal fat balance (FFB) studies: Parents were shown how to complete accurate dietary and pancreatic enzyme records of their child's intake for 5 days, using household measures (including scales where possible) (Appendix 6.3). Parents were advised to collect all stools from their child for 4 days, commencing after completion of the first 24 hours of dietary recording (Appendix 6.3). Detailed information was provided on both these aspects as most parents had not previously completed FFB studies at home and it was important to ensure that standard procedures were followed. Parents were advised that the information they collected would be used to provide feedback regarding fat-based pancreatic enzyme dosing and fat absorption.

The Foodworks computerised food composition database, 1997 (Xyris Software, version 1.05) was used by a dietitian to analyse the dietary records to determine each child's mean intake of fat. The lipid content of the wet stool was measured and expressed as percent faecal fat excretion (%FFE). Percent FFE is the mean daily coefficient of fat absorption (165) and the normal level is considered to be between 0 and 7% (65). Two different criteria, namely >7% and >10% FFE, were used in the analysis as centres differ in what they consider as normal absorption for CF.

PERT doses: Traditionally, the practice at Princess Margaret Hospital was to recommend a weight-based PERT dose of approximately 5 000 IU lipase/kg/day (ie. 1 Pancrease (Janssen-Cilag, 5 000 IU lipase/capsule) or ½ Cotazym-S-Forte (Organon Australia, 10 000 IU lipase/capsule)/kg/day). The dose was increased and adjuvant therapies (eg. a H_2-antagonist) considered if signs of possible malabsorption were evident. Individuals with CF and their parents were advised against self-adjusting the dose in the 1990s when fibrosing colonopathy was identified by other centres.

With the introduction of fat-based dosing at Princess Margaret Hospital, the CF clinic dietitian and gastroenterologist recommended a dose of either 1 250 or 1 700 IU lipase/g of dietary fat to each child, depending on the individual's pre-intervention dietary and pancreatic enzyme records and FFE results. For example, if a dietary record indicated that a child was taking approximately 1 700 IU lipase/g of dietary fat, and their level of FFE was ≤7%, then they were advised to take a lower dose, such as 1 250 IU lipase/g of fat. These doses (1 250 and 1 700 IU lipase/g of fat) corresponded to the more simple expression of one capsule per a specific number of grams of fat, depending on the type of pancreatic enzyme preparation used (ie. a recommended dose of 1 250 IU lipase/g of fat was achieved by taking one Pancrease capsule/4 g of fat or one Cotazym-S-Forte capsule/8 g of fat).

Pre- and post-intervention, the amount of lipase taken with each meal and snack was compared to the child's recommended fat-based dose. Such analyses have not been reported in the literature, so the investigators rated the dose of pancreatic enzymes taken as excessive if the subject took ≥10 000 IU lipase/meal or snack more than his/her recommended dose, and as inadequate if he/she took ≥5 000 IU lipase/meal or snack less than his/her recommended dose. The percentage of meals and snacks, or occasions, accompanied by excessive and inadequate doses of pancreatic enzymes was then calculated. The subject's level of inadequate dosing was further classified as minor or major, depending on both the amount of lipase and percentage of occasions that were under-dosed.

Anthropometry: The children were weighed wearing underclothes on a digital scale (Wedderburn, NSW, Australia) to the nearest 0.1 kg. Stretched stature was measured in triplicate with a stadiometer (Holitan, Crymych, England) to the nearest 0.1 cm and the median value recorded. Weight and height z-scores were determined using the Ozgrow Software Program, 1995 (Camperdown, Australia) which compared the subjects' measurements to the NCHS age- and gender-norms (136).

Confounders: Factors that could influence the effects of PERT (adjuvant therapies to improve the pH of the gastrointestinal tract, gut surgery and tube feeding) were recorded pre- and post-intervention (Appendix 6.4). The methods used to assess the pancreatic enzyme dosage requirements and FFE levels of these subjects were the same as for all other subjects.

Informed consent and ethics approval were not required as this study was primarily an assessment of a change in clinical practice. After providing feedback to parents about their child's results, patient data were collated and analysed anonymously.

Statistical analysis

The Statistical Package for Social Sciences 6.1.3, 1995 (SPSS Incorporated, Chicago, Illinois, USA) was used to analyse differences in pre- to post-intervention measurements by paired t-tests and Wilcoxon matched-pairs signed-ranks test. The associations between %FFE and explanatory variables (eg. amount of lipase, percentage of occasions of inadequate dosing and adherence) were assessed using correlation, linear and logistic regression analysis.

Results

Participants

Complete data were not available on 9 subjects because pre-intervention dietary records were incomplete (n=1) or post-intervention data was not collected (because the child transferred interstate during the intervention period (n=4) or the parents

chose not to complete the second FFB study (n=4)). Thus, the data analysed were of 29 subjects (14 males, 15 females) with a mean age of 7.6 years (range 1.6 to 13.6).

Fat absorption

Subjects were maintained on the same pancreatic enzyme preparation throughout the study. Nineteen subjects took Pancrease (10 males, 9 females) and 10 subjects used Cotazym-S-Forte (4 males, 6 females).

Four subjects had a short-gut and one of these was on long-term nocturnal polymeric gastrostomy feeds. During the study, several subjects were taking an H_2-antagonist (ranitidine) (n=2 pre- and post-intervention), synthetic prostaglandin (misoprostil) (n=3 pre-intervention) or a proton-pump inhibitor (omeprazole) (n=3 post-intervention). The nocturnal gastrostomy feeding regimen was the same both pre- and post-intervention.

Pre-intervention, the dose recommended to 22 subjects was 1 250 IU lipase/g of dietary fat (Pancrease n=17, Cotazym-S-Forte n=5). The remaining seven subjects were prescribed a dose of 1 700 IU lipase/g of dietary fat (Pancrease n=2, Cotazym-S-Forte n=5).

Pre-and post-intervention, the mean level of FFE was 11% (range 2 to 33) and 10.3% (range 4 to 20), respectively. The mean level of FFE of those on Pancrease was not statistically significantly different to those on Cotazym-S-Forte. Table 6.3 shows that the proportion of subjects with normal FFE levels (27% and 25% pre- and post-intervention, respectively) remained relatively constant throughout the study, but the percentage with FFE levels >15% decreased with the intervention. It is interesting to note that only one-third of the sub-group who had FFE levels >15% were children who have short-gut syndrome and that only two of the four subjects with a short-gut had FFE levels >15% pre-intervention.

Table 6.3

Faecal fat excretion (FFE) levels of children with cystic fibrosis
receiving pancreatic enzyme replacement therapy (n=29)

FFE	Percentage of children	
	Pre-intervention	Post-intervention
0 to 7%	27	25
> 7% to ≤ 15%	52	65
> 15%	21	10

Paired t-tests indicated that the mean increase in fat intake, decrease in lipase intake and improvements in %FFE, z-height and z-weight scores were non-significant (p>0.05) (Table 6.4). Comparisons between the individual's recommended dose and their actual dose indicated that the percentage of occasions accompanied by excessive or inadequate amounts of lipase decreased significantly pre- to post-intervention (p=0.001 and p=0.040, respectively) (Table 6.4). Although the percentage of occasions of inadequate dosing improved, the level after the intervention was clinically unacceptable (21%, representing one meal or snack every day).

Table 6.4
Changes in fat and lipase intakes, absorption and anthropometric characteristics of children with cystic fibrosis-related pancreatic insufficiency, aged 1 to 13 years (n=29)

Measure	Unit	Pre-intervention mean±sd	Post-intervention mean±sd	Estimate (95% CI)[e]	P-value
Fat intake	g/day	91±27	94±32	-3 (-9, 4)	0.39
Lipase intake	IU/kg/day[d]	6390±2751	6202±2019	188 (-469, 846)	0.56
	IU/g dietary fat	1532±474	1583±308	-51 (-233, 130)	0.56
Dose excessive[a]	% meals & snacks/day	25±23	9±12	16 (7, 24)	0.001
Dose inadequate[b]	% meals & snacks/day	30±17	21±13	9 (0.4, 17)	0.040
FFE[c]	co-efficient (%)	11.0±6.5	10.3±4.2	0.7 (-1.4, 2.9)	0.500
Height	z-score	-0.5±1.0	-0.4±1.1	-0.1 (-0.1, 0.1)	0.096
Weight	z-score	-0.5±0.9	-0.4±0.9	-0.1 (-0.2, 0.1)	0.250

Paired t-test
[a] ≥ 10 000 IU lipase in excess
[b] ≥ 5 000 IU lipase inadequate
[c] faecal fat excretion, percentage of dietary fat excreted
[d] international units
[e] estimate (confidence interval) for the difference between pre- and post-intervention mean values

Correlation, linear and logistic regression analysis indicated that %FFE was not significantly associated with either the percentage of meals and snacks that were inadequately dosed, the total amount of lipase that was inadequate or the level of inadequate dosing. Table 6.5 shows that subjects classified as having a major level of inadequate dosing (based on the amount of lipase and percentage of occasions of inadequate dosing) were more likely to have abnormal FFE (>7%) than those with minor levels of inadequate dosing, both pre- and post-intervention. The effects of age, gender, type of pancreatic enzyme preparation and time since diagnosis of CF were all non-significant when a multi-variate logistic model was fitted (Table 6.5).

Table 6.5

Effect of a major level of inadequate dosing on faecal fat excretion (FFE)
levels in children with cystic fibrosis-related pancreatic insufficiency (n=29)

	FFE	OR[c]	95% CI[d]	P-value
Pre-intervention	> 7%[a]	3.3	(0.47, 22.61)	0.23
	> 10%[b]	8.2	(0.47, 142.57)	0.15
Post-intervention	> 7%[a]	12.2	(0.83, 178.30)	0.07
	> 10%[b]	1.7	(0.19, 14.62)	0.64

Logistic regression analysis
[a] after adjusting for age, gender and type of enzyme preparation
[b] after adjusting for age, gender, type of enzyme preparation and time since CF was diagnosed
[c] odds ratio
[d] confidence interval for the odds ratio

Four subjects who had pre-intervention enzyme doses greater than the suggested maximum safe level of 10 000 IU lipase/kg/day were able to reduce their intakes of lipase significantly (p=0.027) without detrimental effects on %FFE, z-height or z-weight scores (Table 6.6). Three of these four subjects took medication that affects the pH of the gut throughout the study and one subject also had a short-gut and was on nocturnal gastrostomy feeds.

Table 6.6

Effect of reducing excessive lipase intakes in children with cystic fibrosis-related pancreatic insufficiency (n=4)

Measure	Unit	Pre-intervention mean±sd	Post-intervention mean±sd	Estimate (95% CI)[c]	P-value
Lipase	IU/kg/day[b]	12 125±1 219	9 162±1 828	2963 (640, 5287)	0.027
Height	z-score	-0.8±0.9	-0.9±0.8	0.1 (-0.4, 0.4)	1.0
Weight	z-score	-1.0±0.8	-1.0±0.7	0.0 (-0.8, 0.6)	1.0
FFE[a]	coefficient (%)	11.5±7.3	11.5±5.3	0.0 (-4.7, 4.7)	1.0

Wilcoxon matched-pairs signed-ranks test
[a] faecal fat excretion, percentage of dietary fat excreted
[b] international units
[c] estimate (confidence interval) for the difference between pre- and post-intervention mean values

Apart from fat intake, other nutrients are not presented as the dietary records were unlikely to represent the child's usual intake. Minimal subject burden was a priority during the study due to the demands of usual daily therapies for CF placed on families. Consequently, most FFB studies were conducted at times convenient to the families, namely school holidays and at weekends. Dietary intake at these times of leisure is often different to usual average intake. Also, carers were not specifically directed to maintain the child's usual intake as the focus of this study was the

assessment of fat absorption, not dietary intake. Consequently, it is likely that the child's intake was altered to what carers found easier to record.

Discussion

The results of this study suggested that fat-based dosing has the potential to assist individuals with CF in successfully reducing their intake of pancreatic enzymes without adverse effects. This is in accordance with the Australian PERT Guidelines, which aim to assist individuals with CF in achieving minimum effective daily intakes of lipase and possibly assist the majority of patients with pancreatic insufficiency to achieve normal FFE levels (0% to 7%). Table 6.6 shows that, pre- to post-intervention, there were no adverse effects on %FFE, height or weight when the mean intake of lipase was reduced and the percentage of meals and snacks accompanied by excessive pancreatic enzyme doses more than halved. The ability to achieve minimum effective doses with fat-based dosing, together with a reduction in symptoms associated with malabsorption, were reasons why parents preferred the new method. Most importantly, matching pancreatic enzyme capsules to fat intake enabled four subjects who were taking excessive amounts of lipase (>10 000 IU/kg/day) to successfully reduce their daily intake to safer levels (Table 6.6).

The high level of inadequate dosing limited this study in its ability to indicate the effect of fat-based dosing on fat absorption. Although number of occasions of inadequate dosing improved pre- to post-intervention (Table 6.4), the level remained clinically unacceptable. Abnormal FFE levels (>7%) were found to be more likely in subjects classified as having a major level of inadequate pancreatic enzyme dosing, based on the amount of lipase that was deficient and the number of occasions that this occurred, rather than related to the percentage of occasions of under-dosing alone. The small sample size, and pre-intervention mean level of FFE being near normal (0% to 7%), would also have contributed to the lack of statistically significant improvements in the outcomes measured. However, other findings and observations support the need for further implementation and assessment of fat-based pancreatic enzyme dosing.

Although the pre- and post-intervention group mean FFE levels of the subjects (11.0% and 10.3%, respectively) were close to normal, the range of the values (2% to 33% and 4% to 20%, respectively) highlighted that some children had excessive levels of FFE. In fact, 73% of the subjects had abnormal (>7%) pre-intervention FFE values (Table 6.1) and the majority of subjects who had FFE levels >15% were children without short-gut syndrome. It is important to note that these results may be biased toward better levels, given that the sample of children are likely to be those whose parents endeavour to achieve optimum PERT with their child (as they volunteered to conduct two FFB studies within a six month time period). Further implementation and evaluation of fat-based pancreatic enzyme dosing is supported by the strong preference for the new method among parents of children with CF. Ninety-four percent of parents indicated that they preferred fat-based dosing over the previous method which was a daily weight-based dose (Section 6.1).

This study highlights the need to determine and address reasons for non-adherence, before the effects of the various methods of PERT dosing can be assessed. A major cause of inadequate dosing is likely to have been poor knowledge about the fat content of food and fluids (91, 93, 94). Knowledge deficits became evident when the dietitian provided feedback to parents about the appropriateness of the enzyme doses recorded during the FFB studies. Post-intervention, many parents indicated that they did not realise exactly how high the fat content was of some very high fat foods, such as chocolate, nuts, donuts, 2 minute noodles, takeaway meals, chips and chicken nuggets. This continuing lack of knowledge was not expected as several strategies had been implemented to enhance children's and parents' awareness of the fat content of food and fluids. Parents had received individualised feedback about matching fat intake more appropriately (based on the dietary records completed prior to the intervention) and families had been instructed about how to quantify the amount of fat in food, using resources such as the *Go and Grow with CF* program material, fat counter booklets and labels on food packaging.

It is possible that non-adherence to fat-based doses was a consequence of parents either not referring to the resources or because they were hesitant to change the amount of pancreatic enzymes taken, by redistributing capsules throughout the day or

by varying the dose within and between meals and snacks, without more direct guidance from a health-care provider. Parents' hesitations about changing PERT practices became obvious when a 6 year old boy took the upper bound of his usual snack dose when he ate a large amount (¾ cup) of roasted and choc-coated nuts on two occasions during the post-FFB study. The subject's post-intervention FFE level was excessive (20%), which was not surprising given that fat-based dosing indicated that he may have required three times his usual amount of lipase on both occasions for this type of snack. This example illustrates the importance of objective assessments of nutrition and PERT knowledge until an adequate level is achieved and is successfully translated into practice. Regular education and support of families may assist the process of change by increasing levels of confidence and facilitating the development of necessary skills.

Inadequate supervision during the post-intervention FFB studies was a third reason for inadequate dosing. Unforeseen circumstances, which frequently occur in the clinical setting (eg. holiday outings, parties and parents being required to work unexpectedly), resulted in some children self-managing their dietary intake and fat-based dosing during the FFB study. If children had not yet mastered fat-based dosing then they may have taken their previous usual dose for meals and snacks when they were not being supervised, which may have been inadequate for the amount of fat consumed.

7. CHAPTER SEVEN: DISCUSSION & CONCLUSION

Malnutrition in CF

Malnutrition is a major clinical problem in CF and weight retardation and linear growth failure are common features of the disease. As growth is limited by chronic undernutrition, malnutrition may be more prevalent than is indicated by the typically used measures of height and weight. The aetiology of malnutrition in CF is mostly due to a combination of factors which challenge the maintenance of energy balance, namely increased energy expenditure, increased gastrointestinal losses and decreased oral intake (14, 24, 25).

Poor nutritional status in those with CF can have adverse effects on the lungs, but can also be the consequence of sub-optimal pulmonary function (9). Malnutrition can reduce pulmonary defence mechanisms, decrease exercise tolerance and have detrimental effects on pulmonary muscles. Conversely, recurrent chest infections and lung inflammation can cause increased energy expenditure, anorexia and an imbalance between oxidant injury and antioxidant levels in individuals with CF. A high intake of fat to increase energy intake and supplementation of dietary antioxidants (vitamins A and E) and anti-inflammatory nutrients (n-3 fatty acids) can influence biochemical responses, such that the course of the disease is positively affected (166). The damaging effects on the lung caused by free radicals produced as part of the inflammatory response during chest infections is thought to be reduced when deficiencies of vitamins A and E and essential fatty acids are corrected. Selenium supplementation may also be useful in the treatment of CF as it is closely associated with the antioxidant effects of vitamin E (167). Even lung tissue regeneration may be improved by adequate levels of zinc (which has an established role in wound healing) (167). The assessment of dietary intake of the children with CF during the clinical trial using a FFQ indicated that the mean values of all the nutrients assessed (including zinc) met the RDIs for the normal population.

As the degree of underweight in individuals with CF closely correlates with reduced survival (3), interventions are needed which optimise nutritional status. Health promotion oriented interventions throughout childhood may be advantageous, in terms of both patient well-being and health-care costs, over the use of invasive methods of support (enteral-tube feeding) which are typically used when nutritional status is clearly compromised. A review of the literature, discussions with dietitians in Australian paediatric centres and a needs assessment with children with CF and their carers attending Princess Margaret Hospital for Children supported a new approach to nutrition education with primary school-aged children with CF.

Given that positive health outcomes are more likely if the development of a health prevention program is based on a theoretical model, constructs from the social learning theory (115, 129) were used to develop a behavioural-based CF nutrition program. It was hypothesised that a program that improved knowledge and facilitated the development of appropriate nutrition behaviours and skills in families would subsequently improve oral intake and absorption in children with CF, such that nutritional status, lung function and survival were optimised. The focus of this research was the development and evaluation of the *Go and Grow with CF* nutrition and behaviour intervention program.

Nutritional management of CF

A high energy, high fat, high salt, nutrient dense diet is the general nutritional recommendation for individuals with CF. Dietary studies of children with CF indicate that elevated intakes are rarely met, perhaps due to ongoing anorexia associated with the inflammatory response caused by chest infections and to a lack of interventions which are effective in increasing knowledge, in improving meal-time behaviours and in minimising gastrointestinal losses of individuals with CF.

Traditionally, in most Australian CF Centres, families are counselled on the nutrition and PERT needs of the child at diagnosis and, thereafter, at outpatient clinics or during

hospital admissions. Information is presented in verbal and written form and is mostly directed to parents. The retention of information during these sessions may be limited by a number of factors including the emotional distress that parents experience at diagnosis and the provision of fragmented information as problems arise at busy clinics and during hospitalisation. The cross-sectional, pre-intervention results of the clinical trial indicated that the knowledge of children and carers about the fat content of food was insufficient, the concept of food and drinks having energy (kilojoule) value was poorly understood and children's knowledge about the salt content of food was average. These results supported implementation of a nutrition intervention program which would enhance knowledge.

Nutrition education is more likely to be effective if programs have a theoretical base (116). A theoretical model ensures that the learning opportunities provided by a program incorporate environmental change and establish structures which enable individuals to act on and maintain knowledge, attitudes and behaviours acquired through education (116). An ideal program also needs to include age appropriate strategies. Three CF nutrition-related behavioural programs which were based on social learning theory constructs, namely the parent training approach by Stark et al (109, 110, 125); the counselling strategies developed by Luder (85); and the Family Education Program developed by Bartholomew et al (92, 96, 113) appeared to be limited by numerous deficiencies regarding features recommended for an ideal health education program for children (Section 2.9).

Potential problems with the group sessions developed by Stark et al (109, 110, 125) include time intensiveness for the health-care provider if implementation of the program was to be on a clinic-wide scale, the possibility of minimal effect with children with relatively normal eating behaviours and poor participation rates in the weekly sessions due to the busy schedules of families with a child who has CF.

The counselling strategy developed by Luder (85) is likely to have limited effect with children due to the lack of opportunities for observational learning, such as in group sessions, and non-inclusion of several sources of reinforcement, such as social support in a group format and material rewards. Also, the time intensiveness of counselling sessions would limit application of the approach by Luder (85) in the paediatric clinical setting.

Initially, the home-based program developed by Bartholomew et al (92, 96, 113) appeared useful, but close examination revealed that the nutrition module for children included only two social learning theory constructs; observational learning through modelling and social reinforcement. Sections 2.8 and 2.9 outline the benefits of including several social learning theory constructs and tangible rewards in a behavioural change program for children. Also, the nutrition and PERT information was not comprehensive and did not guide children in making incremental changes to their intake.

Examination of the literature (Section 2) and of the three nutrition-related CF programs (85, 96, 125) (Section 2.10) indicated the need to develop a behavioural-based nutrition prevention program for children with CF. I developed *Go and Grow with CF* in order to address the need for a nutrition and PERT program which incorporated strategies which are known to be effective in changing the health-related behaviours of children (Section 2.9). A clinical trial was then established to assess the effects of *Go and Grow with CF*.

The intervention program

The *Go and Grow with CF* program design was based on the social learning theory of behavioural change (115, 129) and aimed to address the deficiencies identified in other programs. Most of the features identified as being essential for achieving health-related behaviour change in children (Section 2.9) were included in *Go and Grow with CF*. Although the literature highlights the advantages of using a group format for health education (81, 101), CF studies (92, 168) and this research (focus groups and workshops) indicated that a program entirely based on group sessions can be limited by poor attendance. The literature highlights the opportunity that home-based materials provide

for maximising carer involvement in a program for children (122). As there are numerous benefits for children associated with group sessions and with home-based learning, *Go and Grow with CF* consisted of both formats. Other social learning theory-based characteristics of *Go and Grow with CF* that were incorporated in order to maximise knowledge and behavioural change in the children, were the provision of comprehensive nutrition and PERT information (behavioural capability), guidance through incremental steps (self-efficacy) and the use of a structured reward system (reinforcement).

Evaluation of the effects of the intervention

The main research question was: will a nutrition intervention program that is based on a model of behavioural change, and specifically designed for children, facilitate the translation of knowledge into behaviour and action, and thereby improve nutritional status? The secondary research question was: will matching PERT dose to dietary fat intake improve fat absorption in children with CF?

During the clinical trial, process evaluation of the intervention program was conducted and changes in knowledge, behaviour, dietary intake and body composition, using anthropometry, were assessed. The subsequent clinical implementation of the revised program provided the opportunity to conduct further process evaluation and to assess the effects of fat-based PERT dosing on absorption.

Process evaluation

The process evaluation of *Go and Grow with CF* was conducted on both the pilot and revised program. Process evaluation of the clinic-wide implementation of the revised program was beneficial in that the results obtained from the pilot program had limited generalisability. The results of the revised program are more likely to be indicative of what other CF centres who implement *Go and Grow with CF* can expect than the results from the pilot program as participation was not restricted and the program developer did

not interview the carers. Consequently, the carers who provided feedback on the revised program were more likely to have expressed negative comments about *Go and Grow with CF*. The interviewer for process evaluation was a dietitian with CF expertise because, as expected, most carers sought guidance about their child's individual nutrition and PERT needs during the telephone calls. One aspect that would not have been overcome by the clinic-wide implementation of *Go and Grow with CF* is the potential bias caused by carers who enrolled in the clinical trial and who participated in the revised program being particularly interested in nutrition-related aspects of CF.

Participation and completion rates of the revised home-based course were lower than for the pilot program, because the target group included irregular clinic attendees (who attended less than once per year). Many of the children who were not involved in the clinical trial, as they declined the offer to enrol, did not participate in the revised program either. In total, 19 carers (26%) who were potential participants chose not to participate in the revised program. Eight out of the 19 carers who chose not to participate in the revised program had children who were nutritionally at risk and three out of the 19 had children who were referred to the dietitian due to inadequate weight gain within a few months of the conclusion of the program. Busy schedules and non-interest in further education were the two most common reasons for non-participation given by carers during the process evaluation of the revised program. The relationship between non-participation in nutrition education and poor nutritional status needs to be addressed if the effects of a prevention oriented intervention are to be maximised. The issues related to non-participation are likely to be factors which also cause irregular CF clinic attendance, non-acceptance of nutritional problems and non-adherence to treatment recommendations.

A key feature of *Go and Grow with CF*, that was recognised during the pilot study, was its flexibility. This flexibility accommodated different lifestyles, learning styles, medical needs and also minimised competition between the program and a busy family life. The home-based course allowed families to determine the best time of the week for them to

complete the worksheets. The flexibility of the program also ensured that families could take extra time to complete the program if there were delays caused by illness and other commitments. However, if a family does not consider CF nutritional needs to be a high priority then the flexible characteristics of *Go and Grow with CF* may not make a significant contribution to their motivation to fit the program into a busy daily schedule.

Sickness unrelated to CF forced some families participating in the pilot program to interrupt the home-based course. For example, all members except the mother of one family experienced chicken pox during the home-based course. The flexible nature of the program allowed them to recommence a few weeks later, without having missed any information. The families considered the illness as a temporary interruption and completed the home-based course soon after the concluding workshop. This is an important feature of the **Go and Grow with CF** model considering the increased likelihood of illness with a chronic disease.

The necessity of a flexible program was most evident at the time of the concluding workshop of the clinic-wide implementation of *Go and Grow with CF* when only 30% of carers had completed the home-based course. One month later, a further 23% of carers had completed the revised program. A few carers suggested *Go and Grow with CF* could be improved by conducting the program over a greater length of time or by not limiting the amount of time available for completion. However, the 21% of carers who started but did not complete the home-based course, did not progress at all with the activities over the extra month following the concluding workshop. This suggests that extension of the amount of time between the introductory and concluding workshops, to greater than four months, is unlikely to result in a greater proportion of families completing the home-based course. Participation rates for the concluding workshop may possibly be improved by informing carers that non-completion of the home-based course does not exclude attendance at the workshops.

An additional example of the program's flexibility is its use with children who encountered dietary and PERT problems. Three families who had initially decided not to participate in *Go and Grow with CF* were advised to complete the relevant section of the home-based course in isolation to address the specific need of the child. For example, when the problem of poor weight gain was identified during a CF clinic appointment, the dietitian recommended that the child and carer complete the three weekly activities in the energy and fat topic of the home-based course. This illustrates the benefit of the flexibility of the modular format of *Go and Grow with CF* in both preventing and addressing nutrition and PERT problems.

Go and Grow with CF was also useful for educating siblings about the dietary-related treatment needs of the child with CF. This observation became evident during the pilot program when carers reported that siblings were more accepting of apparent diet-related inequalities in the home when they became more knowledgable about the therapeutic needs of a child with CF. Consequently, invitations to the revised *Go and Grow with CF* workshops were extended to all family members. A majority of children attending the revised program workshops were accompanied by a brother or sister, and carers appreciated the opportunity for siblings to learn, directly from health professionals, about the varying dietary requirements of different members of the family. While reviewing the home-based course it also became evident that some children really enjoyed weekly learning sessions involving several members of the family. The flexibility of *Go and Grow with CF* to be used in this way may have provided an opportunity to enhance the learning of children who prefer group-based learning over individual learning.

Finally, the flexibility of *Go and Grow with CF* also meant that the program could be adapted for the small percentage of children with CF who have pancreatic sufficiency. The topics that explore energy and fat, vitamins and minerals, salt and growth are relevant to all children with CF. Children who had pancreatic sufficiency were directed to complete these topics but omit or only read the worksheets on enzymes and malabsorption during the clinic-wide implementation of *Go and Grow with CF*.

Learning in the home environment was another key feature of *Go and Grow with CF* as health behaviour changes of primary school-aged children are maximised by involving their carers (122). Carers reported that they and their children learned how to achieve a high energy, high salt diet and minimise malabsorption by participating in the program. These concepts are fundamental for children with CF and their carers to understand and apply in order to achieve optimum growth and health.

Following is a list of possible reasons why carers and children may have understood concepts more clearly through participation in *Go and Grow with CF*.

1. The *Go and Grow with CF* program provided comprehensive nutrition and PERT information which assisted carers in formulating a structured conceptual framework.

2. The home-based course maximised carer involvement as role models, teachers and reinforcers of behavioural change.

3. The information and activities in *Go and Grow with CF* were in a format appropriate for primary school-aged children.

4. The inclusion of group sessions provided an opportunity for participants to learn from and be motivated by one another to complete the home-based course.

A structured reward system was a very important component of *Go and Grow with CF* as rewards reinforce the behaviour change process for children (124). The aim of the structured reward system was to reinforce desirable nutrition and PERT behaviours being learned through *Go and Grow with CF* in ways most suited to each individual child. As the use of rewards was a new concept for many carers and one not fully understood by others, the introductory workshop provided an avenue for explaining the system. However, some carers needed extra assistance to help identify rewards valued by the child and to ensure that the system was initially used only for behaviours related to the program. A major addition to the revised program was the provision of detailed information about the reward system and steps for establishing it at home. The aim of

this information was to ensure that carers, including those who did not attend the introductory workshop, understood the value of the reward system and were well equipped to implement this approach, which may have been new to some and one not fully understood by others. The dietitian used monthly telephone contact with carers as an avenue for reinforcing the use of the reward system throughout the home-based course.

An important inclusion in the supplement for parents section of the *Go and Grow with CF* home-based course was information on nurturing desirable nutrition and PERT behaviours in children with CF from a very young age in order to reduce the incidence of problems, such as food and enzyme refusal. Some carers may be unaware of appropriate strategies for reinforcing desirable behaviours in children, and others may be tempted to abandon strategies which are effective in the long-term (eg for meeting daily energy requirements) in preference for achieving short-term goals (eg the child eating at each meal and snack). Consequently, *Go and Grow with CF* reinforced that an appropriate strategy for dealing with a child's refusal to eat is to ignore the behaviour and not offer anything more to eat or drink until the next scheduled meal or snack, when the same or an equally nourishing food should be presented. Although carers may be reluctant to deal with their child in this way because of the emphasis placed on adequate growth in CF, they need to be encouraged to avoid inappropriate short-term strategies, such as coaxing their child to eat or preparing a favourite food, in order to prevent long-term meal refusal.

Go and Grow with CF is a nutrition and behaviour program which was highly recommended by families. Similar to the process evaluation of the pilot program, 100% of carers who completed the revised home-based course indicated that they would recommend *Go and Grow with CF* to other families with a child who has CF. Reasons for recommending the program were related to the provision of information explaining what CF nutrition and PERT behaviours are required, the rationale for these requirements and how to successfully accomplish these behaviours. The high level of recommendation of the *Go and Grow with CF* program may also be linked to carers feeling more

confident with the nutritional and PERT care of their child who has CF, as reported during process evaluation.

Successful implementation of *Go and Grow with CF* was dependent on the motivation of the dietitian to conduct the program and adequate time allocation. The time required to conduct the revised *Go and Grow with CF* program at Princess Margaret Hospital averaged at 2.5 hours per child over four months. This was similar to the previous evaluation of the pilot program, except that the time spent implementing the revised program was over a greater length of time as some families did not complete the home-based course until one month after the concluding workshop. This result indicates that expected savings on dietetic time were not achieved by conducting *Go and Grow with CF* with a greater number of participants (nearly two and a half times as many families participated in the revised program compared with the pilot program). However, it is important to note that two new major CF nutrition concepts were introduced in the revised version of *Go and Grow with CF* and much of the dietitians' time was spent reviewing carers' understanding and perceptions of the new guidelines and discussing results from faecal fat balance studies. The time intensiveness of the monthly telephone calls by the dietitian over the 3 months of the intervention program indicates that large clinics may need to acquire additional resources (dietetic time and finance) in order to implement the preventive *Go and Grow with CF* program. Alternatively, *Go and Grow with CF* could be conducted with subgroups of the target group each year in order to decrease the impact of the additional time required by the dietitian to conduct the program. Implementation of an intervention program, such as *Go and Grow with CF*, which includes aspects regarding routine management and anticipatory guidance, as advocated in the CF nutritional management consensus document (18), could be advantageous in streamlining the nutritional management of individuals with CF and in helping families develop the skills necessary for effectively dealing with fundamental nutrition and pancreatic enzyme issues. These outcomes could result in better use of dietetic expertise in the CF clinic in the long-term as the demands of ad hoc problem solving may decrease.

Taking all these factors into account, it is likely that *Go and Grow with CF* will be an efficient and effective method for providing comprehensive, up-to-date dietary and PERT information to families who have a child with CF. Assessment of *Go and Grow with CF* by other CF centres would be beneficial in determining if the program needs any other modifications. Further assessment could include

- ongoing process evaluation
- pre- and post-measurements of dietetic time spent in ad hoc problem solving, and
- an assessment of the effect of the program on fat and energy intake, absorption and the nutritional status in children who have CF.

Knowledge and self-management

Periodic, objective assessment of the knowledge and self-management skills of children with CF and their carers is needed to help identify deficits which need to be addressed. The lack of suitable instruments has limited this process in the past and led to the development of the nutrition and pancreatic enzyme knowledge and self-management questionnaires described in this thesis (Section 3.5).

The cross-sectional assessment indicated that the level of knowledge of children with CF and their carers was higher for PERT than for nutritional aspects. The results highlighted that basic nutritional concepts were not well understood. For example, many respondents did not know that egg is a source of fat. The longitudinal analysis indicated that the *Go and Grow with CF* home-based program was not effective in increasing children's and carers' knowledge and self-management in the long-term. However, the intervention was effective in increasing children's knowledge in the short-term. The apparent absence of a long-term effect of a single exposure to the program, on children's and carers' knowledge, and carers' appropriate management of food and PERT-related behaviour problems, suggests that regular, ongoing education and counselling is required for families in order to reinforce aspects related to the child's current stage of development and disease status. Adequate levels of knowledge are required to enable individuals with

CF to be capable of achieving nutrition and PERT recommendations, and subsequently optimise their nutritional status. It is likely that the deficits in knowledge about dietary fat could limit the ability of children to achieve the high fat diet recommended for CF and to apply fat-based PERT dosing according to the Australian Guidelines.

There was a large difference between the amount of perceived learning, as reported by carers during process evaluation (Sections 5.2 & 6.1), and the objective assessment of knowledge using questionnaires in the clinical trial. This difference may be because the detection of knowledge gains was limited by a number of factors associated with the questionnaire design, even though face, content and consensual validity had been established. The factors that may have limited the knowledge and self-management questionnaires include the following:

- prompting appearing to cause children to increase both the number of appropriate and inappropriate responses to a greater degree than carers,
- reliability testing during piloting not detecting the poor discriminatory power of half the questions that remained in the carers' final knowledge questionnaire, which lead to high baseline rates for carers, and
- the number of questions in each domain with yes and no as the correct response, and with appropriate and inappropriate response options, being disproportionate.

It is possible that the greater proportion of questions in the children's knowledge questionnaire with a positive response being correct gave younger children, who appeared to be guessing 'yes' to many questions, a false high score. The problem of young children possibly guessing answers to questions, and their limited ability to comprehend questions, partly explains why there is a lack of validated nutrition and enzyme questionnaires for this age group. Even though it may be difficult to assess children's knowledge and self-management skills, this process is necessary in order to identify deficits in these areas so that appropriate education can be provided.

There is a growing awareness of the need for children to participate in age-appropriate education programs rather than rely on their parents for information about CF (91, 95, 96,

103, 169). Although children's and carers' scores were statistically significantly correlated, the knowledge score of the children was considerably lower. It is important to maximise the knowledge of children as they are often responsible for their dietary intake and PERT, such as when at school. Also, some carers may expect children to take pancreatic enzymes without reminders, as in this study, from as young as 5 years of age. These findings suggest that CF centres need to guide carers regarding developmentally appropriate levels of self-management for their child. As intended by *Go and Grow with CF*, children could become more cooperative with achieving CF dietary recommendations and appropriate PERT if education programs are age-appropriate and equip them with adequate knowledge, including the reasons for their requirements.

The issue of developmentally appropriate levels of self-management is addressed in the *Go and Grow with CF* program. For example, the activity regarding remembering enzymes directs children and carers to determine together what level of support is needed to optimise pancreatic enzyme administration. The malabsorption module reinforces the need for children to communicate with carers when signs of possible malabsorption occur. *Go and Grow with CF* also endeavours to ensure that families do not become over-confident with aspects of CF care. For example, the enzyme module of the home-based course reminds families about the importance of determining enzyme dosage needs in consultation with CF clinic staff and warns against exceeding their individualised maximum dose (Appendix 3.1, page 8).

The questionnaires used in the clinical trial need to be refined and the effects of guessing minimised by developing an even number of unambiguous questions in each domain with yes and no as the correct response or with appropriate and inappropriate response options. Also, questions should be included to assess knowledge about the fat content of mixed meals, rather than just single foods, to improve the discriminatory power of the questionnaires and determine if respondents can vary pancreatic enzyme doses to match their fat intake appropriately. Reliability and validity of the questionnaires could be improved by evaluating the revised versions with a similar, but larger sample size.

Refinement of the questionnaires, and inclusion of questions with high discriminatory power, would enable the real effects of *Go and Grow with CF* over time, on both children's and carers' knowledge and self-management skills, to be established.

With further refinement, the nutrition and self-management questionnaires for children with CF and their carers, that were developed as part of this research, could become valuable tools in both clinical and research settings. Knowledge deficits could quickly be identified by administering the questionnaires in waiting areas during routine clinic appointments. The dietitian could then schedule a time to discuss responses with the family. The questionnaires could also become valuable research tools for assessing what interventions are needed and in planning and assessing the effects of a program.

Behaviour

The apparent limited effect of the *Go and Grow with CF* intervention program in reducing the frequency of adverse dietary behaviours of the children suggests that either the program had no effect or an instrument designed specifically to assess the behavioural component of the *Go and Grow with CF* program may have been needed. The behavioural section of the *Go and Grow with CF* home-based course supplement for parents may need to be expanded to ensure that the issues assessed by the BPFAS are adequately covered. This research and experience suggest that it is likely that carers will benefit from participation in specific programs and provision of individualised counselling regarding the management of food and PERT refusal, particularly those carers with toddlers and pre-school-aged children with CF who may be dealing with such issues more often than parents of older children. The BPFAS would be useful for ongoing assessment of children with CF, so that behaviour management problems are identified and treated early, before issues escalate (18, 106).

Dietary intake

There are numerous methods available for assessing dietary intake, such as weighed intake records, dietary record diaries, duplicate portions, food frequency questionnaires (FFQs), 24 hour recalls and diet histories. Questionnaire-based methods of dietary assessment are advantageous as they are relatively simple and inexpensive to administer. In comparison to other methods, the specific advantages of the FFQ are that they can be self-administered and are less time consuming for the subject as they do not require food to be weighed. The conclusion to Section 5.5 details why a FFQ was chosen to assess the dietary intakes of the subjects enrolled in the clinical trial. Although the reliability and validity of FFQs are less than optimal, they do provide a measure of habitual intake and have been widely used in studies where the intention was either to describe mean nutrient intake for a group, rank individuals in terms of levels of nutrient intake or to describe patterns of food intake (60).

The GPHNS FFQ was used in the clinical trial to assess the mean change in energy and key nutrient intakes in children with CF, in preference to other FFQs, as its meal-based structure is considered to improve the accuracy of respondent recall of dietary intake, in terms of frequency of consumption and portion size. Furthermore, its comprehensive list of over 300 foods is thought to enhance the accuracy of the nutrient intake data obtained (64). Also, the comprehensive food list of the GPHNS FFQ was considered by the investigators to be advantageous as it included many of the wide range of foods from which children with CF are typically encouraged to select, in order to achieve their high energy and high salt dietary requirements, and would, thus, reflect energy intake more accurately than other types of FFQs, which do not list such foods. An additional important feature of the GPHNS FFQ was the inclusion of serve size specifications, which appeared to the investigators to be appropriate for children with CF. These serve size specifications enabled quantitative changes in nutrient consumption to be assessed. A final factor which was taken into consideration was that software for nutrient analysis of the GPHNS FFQ was readily available.

The apparent lack of an effect of the *Go and Grow with CF* program on the dietary intake of children with CF in this study may be due to the true lack of change or possibly due to factors associated with reporting intake or specifically due to the use of a FFQ. Nutrient intakes, especially those contained in fruit and vegetables, have been found to be over-estimated using a FFQ (62-64, 152-154). This research suggests that FFQs should not be used to assess a group of children with CF as dietary intakes appeared to be highly overestimated when carers completed the questionnaires on behalf of the children, as discussed in Section 5.5. An alternative method for assessing the dietary intake of children with CF, without requiring too much effort from the respondent, could be multiple days of unannounced 24-hour recalls (170). Bingham et al (156) support the use of the 24-hour recalls with the general population as they found nutrient intakes obtained from 50 to 65 year old women using this method compared favourably with weighed records. However, it is possible that 24-hour recalls may be inappropriate for a longitudinal study as it may be difficult to differentiate between short-term variability and long-term change in nutrient intakes (171). Consequently, dietary records may need to be used, particularly if further evaluation of the revised *Go and Grow with CF* program is to include an assessment of fat absorption through faecal fat balance studies.

Body composition

Assessment of nutritional status typically involves the interpretation of information obtained from dietary, biochemical, body composition and clinical studies. Numerous factors affect the reliability and validity of methods used to determine nutritional status, such that there are no gold standards. This may partly explain the continuing heavy reliance on measures of height and weight for the assessment of nutritional status in children with CF in both the clinical and research setting.

The extensive anthropometric data obtained during this study suggest that measurements of height and weight may be misleading when used to determine nutritional status. The subjects were thought to be reasonably well nourished, as they were without secondary conditions known to adversely affect nutritional status (short-gut syndrome and liver

disease) and the mean %IBW values for both genders were within the normal range for CF (90% to 110% IBW) (18). However, comparisons of skinfold and girth data with a reference group (143) suggested that there was a large deficit in the muscularity of the children with CF, indicative of malnutrition.

The %IBW index may be useful for distinguishing those who are thin or heavy for height from those who are well proportioned, with regard to height and weight. But the %IBW index does not detect growth deficits relative to an individual's genetic potential for height. Also, malnutrition is often underestimated when the %IBW index is used as individuals who are short and thin are classified as normal. Deteriorations in nutritional status will not be detected by the %IBW index if reductions in height and weight gain occur at similar rates. Conversion of height and weight measurements to z-scores is more beneficial than the %IBW index as the height and weight comparisons made with a reference group are separate.

Preferably, assessments of nutritional status in individuals with CF should include body composition analysis in order to determine important information about the energy stores and protein reserves of the body. Deterioration in nutritional status could be detected early by monitoring changes in body composition, prior to weight gains being inadequate and growth being stunted (13, 18, 52). Routine longitudinal monitoring of fat and fat free mass in CF could be used to assess the efficacy of medical and nutritional therapies, in maintaining or improving nutritional status and growth parameters. Also, the effects of nutritional status on morbidity and mortality associated with the disease could be monitored.

The higher mortality rate for females with CF and the lower mean age of survival is poorly understood; it is possible that the aetiology is linked to nutritional status. Rosenfeld et al (17) suggest that pre-pubertal malnutrition and essential fatty acid deficiencies may be partly responsible for the worse survival rate among younger females with CF. The differences in the body composition of pre-pubertal males and females

with CF in the clinical trial, prior to the intervention, support further investigation of the relationship between body composition and gender differences in lung disease and survival. Although the mean age- and gender-standardised anthropometric values were similar between genders for the 10 and 11 year old children with CF, the values of the younger females (2 to 7 year olds) were lower than the males. These gender differences may well be related to differences in dietary intake. Collins et al (172) suggest that the lower dietary intake of fat and energy by females with CF than males may possibly limit the beneficial effects of diet on nutritional status and of dietary essential fatty acids and antioxidant intake on lung disease. The mean age and dietary intakes of the females and males were 11.2 and 13.2 years, 83.9 and 128.2 g fat/day (p=0.004) and 106.7 and 131.2 %RDI for energy (p=0.028), respectively (172). Future investigations regarding the relationship between gender, malnutrition and survival will require comprehensive assessments of nutritional status.

The choice of which body composition method to use is not well defined as a number of factors (accuracy, precision, cost, ethical rating, technical difficulty, portability and subject acceptability) influence the type of method that is most suitable. The difficulties experienced in assessing the body composition of children are due to impractical issues associated with many of the standard methods and many adult-based assumptions (eg. constant fat free mass composition) are not valid in children. Furthermore, the assessment of body composition in CF may be complicated by factors related to lung disease. The main techniques available for use with children, which are non-invasive and have low radiation exposure, include densitometry, skinfolds, bielectrical impedance analysis, dual energy x-ray absorptiometry, in vivo neutron activation analysis, total body potassium and total body water. The methods which rely on a two component (fat and fat free mass) body composition model (eg. skinfolds) are less accurate in children as their fat free mass is continually changing. Multi-component (fat, muscle, bone) models (eg. dual energy x-ray absorptiometry) provide the most accurate and precise assessment of body composition in children because there are fewer assumptions. However, factors

such as ethics, subject acceptability, cost, difficulty and portability cause multi-component models to be less preferred.

As the potential usefulness of extensive anthropometry in the CF population has not yet been examined, one aspect of the research project was to determine if a range of anthropometric measurements could detect early signs of malnutrition in pre-pubertal children with CF, in addition to assessing the effects of the *Go and Grow with CF* intervention program on the body composition of children with CF.

Anthropometrists advocate a range of sites be measured to enhance the validity of anthropometric assessments of body composition (Section 2.5). Based on our finding in the cross-sectional analysis of a large deficit in the muscularity of children with CF, we also recommend that a large range of sites should be measured when anthropometry is used to assess the nutritional status of this population. This recommendation is supported by the longitudinal study by Stettler et al (2) which illustrated the limitations of relying on one site to indicate changes in fat free mass. In the study by Stettler (2) the mean upper-arm muscle area of the children with CF, as determined by anthropometry, was not statistically significantly different to that of the control group, but fat-free mass of the boys with CF, as determined by skinfolds from four sites, total body water and total body electrical conductivity, was lower. Monitoring of measures of body composition, by graphing serial z-scores of the anthropometric measures or by using a computer program to display comparisons between different occasions, could be used to assess the effects of various interventions. An example of a tool for monitoring is the O-Scale System for Assessment of Human Body Composition 1985 (Ross & Ward, Surrey, B.C.:Rosscraft) computer program which relates individual height-corrected anthropometric data to the COGRO age- and gender-norms and produces adiposity and proportional weight ratings, raw scores and a height-adjusted physique profile.

While the impact of the intervention on children's health status was assessed using measures of anthropometry and pulmonary function, the clinical study did not detect a

statistically significant improvement in nutritional status either because the intervention had no effect or that the study was limited by factors related to a small sample size, short duration of the intervention and of follow-up, the effects of bouts of infection and concurrent illnesses. We were limited by a clinic population of only 70 eligible subjects which was then reduced to 59 subjects who were willing and able to enrol. A significant characteristic regarding many of the children whose carers declined the offer to enrol in the clinical trial was their poor nutritional status. The bias toward the better nourished children with CF being enrolled in the clinical trial suggests that it is more likely that the outcome that could be anticipated from a prevention program like *Go and Grow with CF* is maintenance of baseline functioning or a slowed rate of decline in intervention subjects. For improvements in the height and weight and anthropometric measures of body composition to be observed in the children with CF, in whom the disease and degree of malnutrition were mild, a larger sample size and longer period of time of follow-up may have been needed to detect positive changes in nutritional status. Studies that have shown improvements in height and weight and pulmonary function with enteral-feeding (19, 20, 51, 77, 78) have typically involved interventions with patients with more severe malnutrition than those enrolled in our study. The benefits of our preventive nutritional intervention on pulmonary function may be most apparent in long-term follow-up when subjects are compared with patients who never participated in the program.

Pancreatic enzyme replacement therapy

Continual advances in pancreatic enzyme preparations and adjuvant therapies have contributed to significant improvements in the nutritional status of individuals with CF over the last two decades. Determination of the most efficacious method of PERT dosing, together with behavioural-based nutrition prevention programs, may lead to further enhancement of the nutritional status and survival of individuals with CF.

PERT doses have traditionally been based on body weight and, as pancreatic enzyme preparations were initially thought to be without adverse effects, the amount was

increased until a reduction in symptoms associated with malabsorption (abdominal pain and loose stools) was achieved (69, 173, 174). The incidence of fibrosing colonopathy, associated with high doses of pancreatic enzymes and/or the use of high strength preparations in the early 1990s (175-177), caused many CF centres to review the practices of their patients and collaborate to produce national guidelines (67, 69, 70). It was likely that some children with CF would be able to reduce the amount of pancreatic enzymes taken without deterioration in fat absorption based on a study by Robinson and Sly (178) which investigated the effects of reducing high intakes.

The assessment of the effect of fat-based dosing (in accordance with the Australian PERT Guidelines) as part of this thesis highlighted the importance of objectively ensuring that PERT doses are adhered to before attempting to draw conclusions about effectiveness. Based on our observations, the assessment of absorption should be delayed until high levels of adherence to PERT doses are achieved. Completion of a series of dietary records of the child's intake would provide a means for parents and children to self-monitor the appropriateness of PERT doses and for evaluation with the dietitian.

Given the high level of non-adherence to fat-based doses during the clinical implementation of *Go and Grow with CF* and the numerous daily demands placed on children and their parents to conduct CF-related therapies (chest treatments, medication and diet), fat-based dosing may be considered as requiring too much effort to be warranted. Also, the fact that PERT requirements of patients with CF-related pancreatic insufficiency vary considerably, within and between individuals due to numerous patient characteristics and the bioavailability of preparations (Section 2.4), may suggest that it is not possible to determine efficacious dosing. However, fat-based dosing warrants further investigation into its effects on absorption, given, firstly, that parents have a strong preference for this method (Section 6.1) and, secondly, by the high prevalence of above normal FFE levels despite advances in the treatment of CF, including the use of adjuvant therapies (approximately three-quarters of the subjects had FFE levels >7%) (Table 6.3). If the variability of the factors which affect digestion and absorption in individuals with

CF is minimal then it may be possible that fat-based dosing can cause significant improvements in fat absorption, particularly in those with elevated levels of FFE. It is likely that the time taken to determine the number of pancreatic enzyme capsules required when using the fat-based method will decrease considerably once families are familiar with the fat content of the child's usual meals and snacks (which are often similar from one day to the next). Improved levels of knowledge about the composition of food may be advantageous in assisting children to achieve the high energy, high fat diet recommended for individuals with CF. Murphy and Wootton (37) support the pursuit of the most effective method of PERT dosing by suggesting that interventions which address the adverse effects of malabsorption-related abdominal discomfort on appetite will enhance the oral intakes of children with CF.

Summary of the evaluation

Similar to the process evaluation of the pilot program, 100% of carers who completed the revised home-based course indicated that they would recommend *Go and Grow with CF* to other families with a child who has CF. Carers' reasons for recommending the program were related to the provision of information explaining what CF nutrition and PERT behaviours are required, the rationale for these requirements and how to successfully accomplish the behaviours. It may be that *Go and Grow with CF* gave carers and children a sense of confidence and of control over the disease through mastery of some of the treatment skills.

Focus groups and the goals carers set for themselves and their children prior to the intervention revealed that carers were very interested in learning and process evaluation of the pilot and revised *Go and Grow with CF* programs revealed a high level of perceived learning by carers and children. Carers also indicated that they were confident about the nutrition and PERT management of their child with CF. The social learning theory model of behavioural change indicates that knowledge needs to be optimal in order for individuals to be capable of performing desired behaviours (115). Increased

confidence is likely to have had a positive effect on carers' self-efficacy, causing desirable behaviours to be more achievable.

The objective assessment of knowledge, using the questionnaires for children and carers that were developed and validated as part of this thesis, indicated a significant improvement in children's knowledge in the short-term (Table 5.5). The lack of significant improvements in any of the parameters other than children's knowledge in the short-term does not support the high level of learning that was perceived by carers. It may be that a once-off implementation of *Go and Grow with CF* alone is insufficient for achieving behavioural change, particularly if inappropriate behaviours have been practised for several years, the child's overall medical treatment is complex and knowledge is poor, as detailed in Section 2.7. Intensive behavioural therapy on an individual basis, before or after a family has participated in a clinic-wide global program, such as *Go and Grow with CF*, may be necessary. Alternatively, a preventive approach to nutrition in CF in the form of participation in several behavioural-based programs throughout the child's life-cycle (at diagnosis, one year post-diagnosis, 2 to 5 years old, 6 to 11 years old, 12 to 16 years old and 17 years and over) may be more advantageous. For patients with complex treatment regimens, there may need to be a high level of involvement of children and/or parents in determining, monitoring and modifying CF therapies, in collaboration with health-care providers, in order for nutrition programs to be effective. Families with poor CF nutrition knowledge may need repeated learning opportunities to reinforce aspects learned in a nutrition program.

This research may have been limited by factors such as the parameters measured and the inadequacies in the instruments used (particularly the questionnaires), the small sample size (which was determined by the relatively small CF population available) and the short duration of the intervention and follow-up. The lack of a statistically significant improvement in the anthropometric measurements with the intervention may also be related to non-participation of children with poor nutritional status. For improvements in the height and weight and anthropometric measures of body composition to be observed

in the children with CF, in whom the disease and degree of malnutrition were mild, a larger sample size and longer period of time of follow-up may be needed to detect positive changes in nutritional status.

Carers' unanimous recommendation of *Go and Grow with CF*, together with high levels of perceived learning and children's increase in knowledge in the short-term, indicate the benefits of the program and support the need for further evaluation using refined and sensitive instruments, with a larger sample size and over a longer period of time. Process evaluation with subjects who are unfamiliar with the program developer is also needed in order to control for a possible clinician effect during this research.

The observation of a high level of non-adherence to fat-based PERT doses highlights the importance of objectively ensuring adherence. When adherence is optimal then the effects of the Australian PERT Guidelines on absorption can be determined.

Recommendation- CF nutrition education

The obvious conclusion from my research is that CF nutritional care should ideally involve a series of programs to address dietary- and PERT-related issues as they relate to the child's stage of development. The optimum time for commencing nutrition programs may be in the pre-school years, when families are faced with fewer competing demands, as the most frequently cited reason for refusal to participate in the programs by carers of school-aged children was lack of time. This research (focus groups and process evaluation) suggested that another reason why participation rates may be higher with carers of younger children is that interest in learning is often highest soon after diagnosis, which most often occurs in the first few years of life and within the first few months of life in populations where neonatal screening for CF is available. Intensive training of parents of pre-school-aged children is reinforced by Stark et al (168) who reported better recruitment rates for a nutrition-related behavioural program with parents of pre-school-aged children with CF compared with school-aged children with CF (80% and 50%, respectively). Conducting several interventions during the pre-school period may be

highly beneficial in that the educational messages can be pro-active and motivational, so that families are encouraged to periodically participate in programs throughout the child's life.

Optimising nutritional status from infancy and throughout childhood, through the use of prevention programs, may ensure that the effects of malnutrition on airway growth are minimised (162). The likelihood of early learning sessions being limited by the emotional distress that parents experience at diagnosis (99) and the need to provide children with developmentally appropriate health-education experiences (101) justify the implementation of programs (such as *Go and Grow with CF*) with primary school-aged children with CF. Clinic-wide programs could be supplemented at each stage of development as necessary by material directed at particular subgroups of the CF population, such as nutrition and body image for pubertal and post-pubertal females. The concept of a series of programs related to the child's stage of development is supported by McKelvey and Borgersen (101) who advocate approaching self-management training of families with a child who has a chronic illness according to the family life-cycle (infancy, toddlerhood, primary school-aged and adolescence).

The outcomes of this research and the findings of Crist et al (106), which indicated that behavioural issues may impact on the dietary intake of many children who have CF, suggest that programs conducted for carers of pre-school-aged children should address the management of child behaviours in addition to providing nutrition and PERT information. Parents of children with a chronic illness like CF face the enormous task of managing behaviours related to the numerous treatments for the disease, in addition to normal child behaviours. Specifically designed parenting programs may help parents distinguish between strategies that enhance appropriate behaviours in children in the long-term (eg. a high energy intake every day) as opposed to achieving desired behaviours in the short-term (eg. consuming a particular meal). Carers of both pre-school-aged and primary school-aged children should also be encouraged to optimise communication with their child about continual monitoring of malabsorption, so that

immediate action can be implemented to overcome the problem. Programs which address normal parenting strategies at each stage of the child's development may help ensure that parental reactions to issues, such as food refusal which may be partly due to anorexia associated with the disease, do not compound the problems.

Children themselves need to participate in nutrition programs on several occasions as their levels of comprehension will increase significantly as they grow older. *Go and Grow with CF* was initially designed for primary school-aged children and carers of 2 to 11 year olds. The intention was to implement the program approximately every four years to reach children during both these stages and to provide savings on the dietitian's time, rather than conduct *Go and Grow with CF* every year with a portion of the target group. Time spent preparing for the workshops, newsletters and evaluation would be spread across a larger number of participants. It is recommended that clinics with a large population conduct several introductory and concluding workshops (with ten to 15 families per workshop) to ensure that group size promotes interaction.

This research supports the development of programs specifically designed for upper primary school-aged and adolescent children. Given that process evaluation of *Go and Grow with CF* indicated that carers of pre-school-aged children needed assistance with adapting the program for their families, the younger age group may also benefit from a specific program. The particular needs of families with a child diagnosed with CF through neonatal screening will also need to be addressed at each stage of development. Colouring books, stories, nutrition games, high energy tasting sessions and computer assisted learning packages could be useful media to use with older children, but the development of a program should be based on a theoretical model of behavioural change and incorporate strategies that are most suited to the target age group. The *Go and Grow with CF* program, which included group and home-based learning, carer involvement, comprehensive information, incremental steps and reinforcement, would be a useful basis for developing learning activities for other programs.

The benefits of optimising nutrition and PERT knowledge and behaviours through the use of *Go and Grow with CF* during childhood, when enthusiasm and motivation to learn are most likely to peak, may be recognised at adolescence. During adolescence, individuals with CF often neglect their treatments, perhaps initially because they do not fully understand how to manage all the aspects. The benefits of effective pre-adolescent training, such as through participation in *Go and Grow with CF* when primary school-aged, may be realised when children become autonomous from their parents and have sufficient skills to maintain optimal management of their disease (82).

Implementation of nutrition-related behavioural programs, at several stages throughout a child's life, may require health-care providers to make major adjustments in their approach to caring for individuals with CF. Resources may need to be reallocated so that there is sufficient time available for conducting programs separate to clinic sessions. Team support for a preventive approach to nutrition will also be needed as families are influenced by the attitude of other professionals towards extracurricular activities. This research indicated that an important source of motivation for carers participating in the home-based course was the interest in the *Go and Grow with CF* program expressed by CF team members (doctors, nurses, physiotherapists).

Recommendation- nutritional assessment

Skinfold and girth measurements are useful for estimating body composition as information is provided about nutritional stores (energy in fat and protein in lean body mass). This research suggests that an extensive range of anthropometric measurements should be taken from children with CF, probably on a yearly basis, if superior methods of body composition assessment are not available or if a non-invasive, inexpensive, portable and relatively quick method is preferred. It would be necessary to measure numerous anthropometric sites as no single measure fully reflects nutritional status (179). Assessments of body composition of children with CF could be advantageous for optimising health outcomes, so that quality of life is enhanced and the costs of invasive nutrition interventions reduced. Individualised feedback based on the child's

anthropometric profile (eg from the O-Scale printouts) could be used as an educational tool during nutrition counselling sessions. Secondly, deteriorations in muscle and fat stores could be detected early, and interventions commenced, before growth and pulmonary status are adversely affected. Depletion of the fat compartment as a consequence of energy insufficiency may be indicative of a child with CF who is more at risk than others for pulmonary exacerbations or other complications of the disease (23). Maintenance of optimum nutritional status, through early detection and treatment of deteriorations, throughout childhood and puberty could also have significant positive effects on survival and possibly reduce the need for and subsequent cost of nutritional support (enteral-feeding) (179). Future validation of skinfolds and girths in children with CF, against superior methods such as dual-energy x-ray absorptiometry, may indicate the particular sites that are most representative of nutritional status. The use of three body compartment models (dual energy x-ray absorptiometry) provides more precise information than two body compartment models (skinfolds) which can only provide an estimate of fat mass and fat free mass.

Recommendation- pancreatic enzyme replacement therapy

The study of the effects of fat-based PERT dosing on absorption in children with CF when *Go and Grow with CF* was implemented on a clinic-wide basis was limited by the high level of non-adherence. This observation suggests that future evaluations of all the PERT dosing methods are needed and should include assessments of dose on a meal and snack basis, rather than just on daily intake, in order for levels of adherence to be examined (39, 164). With regard to implementation of fat-based dosing, this research suggests that individuals with CF or their parents should complete a series of detailed 3 to 5 day dietary and pancreatic enzyme records (Appendix 7.1) until a high level of adherence is achieved, such as no more than one meal or snack per day being under-dosed by 5 000 IU of lipase or less. This process of self-monitoring may become an important factor in assisting individuals with CF in changing their pancreatic enzyme dosing practices. The dietary and pancreatic enzyme records will also enable the dietitian to provide periodic, individualised, objective feedback about the appropriateness of

pancreatic enzyme doses. Collaborative research between CF centres could provide much insight into PERT dosing methods. Fat-based dosing could potentially influence the degree of maldigestion and malabsorption in individuals and suggest whether or not the level is constant, taking the varying levels of enzyme activity in preparations into account. Studies based on the recommendations made from this thesis are needed to establish evidence-based PERT dosing practices and to ascertain if fat absorption can be improved to near normal levels (0 to 7% FFE) in the majority of individuals with CF. Studies are also needed to determine optimum PERT dosing for enteral-feeding of polymeric preparations, including both timing of administration of pancreatic enzymes and dosage (68). Therapeutic improvements in fat absorption could lead to further enhancement of nutritional status, lung function and survival in individuals with CF.

Conclusion

The key to reducing morbidity and mortality and in enhancing the life expectancy of individuals with CF may be in preventing malnutrition. Interventions which at least minimise or delay deteriorations in nutritional status may have positive effects on growth, physical activity, pulmonary function, quality of life and survival. Comprehensive nutrition prevention programs, such as *Go and Grow with CF*, may be a more cost-effective means for addressing nutrition than the use of more invasive treatments (enteral-feeding) when nutritional status is clearly compromised. Nutrition interventions in CF may be most effective when implemented in early childhood in order to maximise the impact of optimal nutritional status on lung development. Periodic participation in behavioural-based educational programs, which are accompanied by appropriate parenting programs throughout childhood, together with adolescent and adult education programs, may result in the life-long maintenance of optimal nutritional status.

The features of *Go and Grow with CF* which are most likely to have made the program suitable for facilitating nutrition and PERT behavioural change in primary school-aged children with CF were: that it was based on social learning theory constructs, namely behavioural capability (comprehensive information), self-efficacy (incremental changes

to meals, snacks and PERT) and reinforcement (a structured reward system); the inclusion of workshops at the beginning and at the end of the home-based course in order to provide opportunities for role modelling and peer reinforcement; learning in the home environment; flexibility of the home-based course; and a structured, integrated system of reinforcing behaviour change. The *Go and Grow with CF* program, which was specifically designed for primary school-aged children, is an ideal model on which to base the development of nutrition programs for other age groups.

The lack of a significant improvement in nutritional status during the clinical trial may have been because either the single exposure to the intervention program had no effect, the parameters measured or the tools used to measure change in knowledge, self-management, behaviour, dietary intake and body composition were not sufficiently sensitive or that the time period of the intervention program and follow-up of parameters were too short.

The high level of perceived learning, improvement in children's knowledge, reported increase in confidence, together with carers' unanimous recommendation of both the pilot and revised *Go and Grow with CF* to other families, highlight the potential usefulness of the program for patients and dietitans in other CF centres. The strategies used during the development of *Go and Grow with CF* are applicable to other aspects of CF care and to other chronic conditions.

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Appendix 3.1

The Revised Intervention Program

- Table of contents

- PERT module

'Go and Grow with CF' is a nutrition and enzyme program for you and your mum or dad to complete together. The 6 topics and 11 activities included in the program are listed below.

Each unit is divided into steps to help you move **from KNOWING to DOING** what is needed with food, enzymes and malabsorption.

Watch for the steps each week. They are:

from KNOWING
1. Plan
2. Do It
3. Feedback
4. Practise
5. Keep Going
6. Get There

to DOING !

Appendix 3.2

Children's Questionnaires

Knowledge and Self-management

Note: For copyright reasons Appendix 3.2 has not been reproduced.

(Co-ordinator, ADT Project (Retrospective), Curtin University of Technology, 9.7.03)

Appendix 3.3

Carers' Questionnaires

Knowledge and Self-management

Note: For copyright reasons Appendix 3.3 has not been reproduced.

(Co-ordinator, ADT Project (Retrospective), Curtin University of Technology, 9.7.03)

Appendix 4.1

Process Evaluation Data Recording Sheets

for the Pilot Program

Since the beginning of this program I last phoned:

Name	Date	What week are you currently doing (1-10)	What new thing have you learned from GO & GROW WITH CF	What new thing has your child learned from GO & GROW WITH CF	What have you enjoyed in the GO AND GROW WITH CF?

Since the beginning of this program I last phoned:

What has your child enjoyed in GO & GROW WITH CF?	What have you not liked in GO & GROW WITH CF?	What has your child not liked in GO & GROW WITH CF?	Are you using the reward system every week. If yes, what rewards has your child chosen. If no, explore reasons why not	Have you or your child identified any nutrition or enzyme problems? If yes, what?. If no, remind the parent about the steps for problem solving sheet if they do	If the family have identified a problem go through the steps for problem solving sheet with them. What was the problem	For children 4 years old or more - does your child swallow enzyme capsules whole? If no, discuss plans for learning to swallow capsules	Have you put into action any of the information from the supplement for Parents section - what to do if - ? If yes: what:

Since the beginning of this program I last phoned:

What have you found difficult about the GO & GROW WITH CF program?	What has your child found difficult about the program?	What improvements do you suggest to the GO & GROW WITH CF program (what could make it better?)	Describe how you felt about the program before you started 1.v. uninterested 2.uninterested 3.indifferent 4.interested 5.v. interested	What is your level of motivation to keep doing the program. 1.v. unmotivated 2.unmotivated 3.indifferent 4.motivated 5.v. motivated	If 1, 2 or 3 Why?	If 1, 2 or 3 What could improve your motivation?	What do you think is your child's level of motivation to keep doing the program. 1.v. unmotivated 2.unmotivated 3.indifferent 4.motivated 5.v. motivated	If 1, 2 or 3 Why?	If 1, 2 or 3 What could improve your child's motivation?

Since the beginning of this program I last phoned:

Prior to the program, you said your program goal for yourself was:	Post-intervention Was this achieved 1 = Yes 0 = No	Prior to the program, you said your program goal for yourself was:	Post-intervention Was this achieved 1 = Yes 0 = No

Since the beginning of this program I last phoned:

What have you found difficult about the GO & GROW WITH CF program?	What has your child found difficult about the program?	What improvements do you suggest to the GO & GROW WITH CF program (what could make it better?)	Would you recommend the GO & GROW with CF program to other parents who have a child who has CF? 1=yes 0=no 9=Don't know/maybe Why?	What reinforces/supports from CF clinic staff/dietitian worked best to keep you on track with the program? 1. Phone call 2. Newsletters 3. Conversations at clinic 4. Other, specify	What reinforces/supports from CF clinic staff/dietitian did you not like? 1. Phone call 2. Newsletters 3. Conversations at clinic 4. Other, specify	What things were good to go over again in the new GO & GROW WITH CF nutrition program?

Appendix 4.2

Modified Behavioural Paediatrics

Feeding Assessment Scale

Appendix 4.3

Food Frequency Questionnaire

- **Additional questions**

- **Instructions**

Appendix 4.4

Anthropometry Recording Sheet

BASIC ANTHROPOMETRIC PROFORMA

File No
Person
Gender
Group

DOB
Date

day   month   year

A   01 Subject ....

B   02 Body mass ....
    03 Stature (stretched)   19
                        23

C   04 Triceps sf   27
    05 Subscapular sf   30
    06 Biceps sf   33
    07 Iliac crest sf   36
    08 Supraspinale sf   39
    09 Abdominal sf   42
    10 Front thigh sf   45
    11 Medial calf sf   48

E   12 Arm girth relaxed   51
    13 Arm girth flexed and tensed   54
    14 Forearm girth (max, relaxed)   57
    15 Wrist girth (distal styloid)   60
    16 Chest girth (mesosternale)   63
    17 Waist girth (min.)   67
    18 Gluteal girth (max.)   71
    19 Thigh girth (1cm dist. glut.line)   75
    20 Calf girth (max)   77
    21 Ankle girth   81

CF NUTRITION STUDY

Appendix 4.5

Household Record Form

File No
Person
Gender
Group

DOB
Date

day   month   year

.... SEIFA
(office use only)

This section is a HOUSEHOLD RECORD FORM which will enable us to compare our study with other studies. All information will again be treated as confidential.

What is your postcode ....

Person No.	A Name	B Relationship		C Sex	D Age	E Date of Birth
	What are the names of the people who live in your home? INTERVIEWER: Please ensure primary carer is 01. Please ensure secondary carer is 02. Primary carer is the person who mostly watches what the child with CF eats and gives out enzymes. Secondary carer is the person who next most often does these things. List children in order of oldest to youngest. Eldest child is 03. Surname Other	What is .... relationship to you? 1. Partner/spouse of person 1 2. Other adult relative 3. Child of person 1 & 2 4. Child of person 1 only 5. Child of person 2 only 6. Foster child 7. Parents of person 1 or 2 8. Other	Are there any step relationships within the household? Yes or No	M=1 F=2	What was age last birth day?	What was date of birth? Day Month Year
01						
02						
03						
04						
05						
06						
07						
08						
09						
10						

HOUSEHOLD RECORD FORM

Person No.	F Marital Status What is .... marital status? 1. In first marriage 2. Remarried 3. Separated but not divorced 4. Divorced 5. Widowed 6. Never married 7. De facto	G. Country of Birth In which country was ..... born? 1. Australia → 2. UK 3. Italy 4. NZ 5. Netherlands 6. India 7. Germany 8. Other (specify) →I	H State In which state was ...... born? 1. NSW 2. VIC 3. QLD 4. WA 5. SA 6. NT 7. ACT 8. TAS FINISHED	I Year Arrived In what year did ..... arrive in Australia?	J Has CF Does .... have CF? Yes or No
01					
02					
03					
04					
05					
06					
07					
08					
09					
10					

J. **What was the highest grade or year you completed at school ?**

Primary school ☐
Year 8 ☐
Year 9 ☐
Year 10 ☐
Year 11 ☐
Year 12 ☐
Don't Know ☐
Other (specify) Level _____
State _____
Country _____

HOUSEHOLD RECORD FORM

K. **What was the highest grade or year completed at school by the secondary carer of your child with CF?**

Primary school	❏
Year 8	❏
Year 9	❏
Year 10	❏
Year 11	❏
Year 12	❏
Don't Know	❏

Other (specify) Level _____

State _____

Country _____

L. **What is the highest qualification you, the primary carer, obtained since leaving primary or high school?**

No post-school	❏
Trade/apprenticeship	❏
Certificate from college, TAFE	❏
Diploma (beyond Year 12)	❏
Bachelors degree	❏
Postgraduate diploma/higher degree	❏
Other (specify) _____	❏
Don't Know	❏

M. **What is the highest qualification the secondary carer of your child with CF obtained since leaving primary or high school?**

No post-school	❏
Trade/apprenticeship	❏
Certificate from college, TAFE	❏
Diploma (beyond Year 12)	❏
Bachelors degree	❏
Postgraduate diploma/higher degree	❏
Other (specify) _____	❏
Don't Know	❏

N. **What was your income before tax per week or per year in the financial year 1994-95?**
- include family allowance and other benefits, child support, superannuation, wages and salaries, overtime, dividends, business income, interest.
- do not deduct tax, superannuation payments, health insurance payments.

Interviewer: Tick one appropriate category

Unprompted

A.	<$58 per week (<$3001 per year)	❏
B.	$58 - $96 per week ($3001 - $5000 per year)	❏
C.	$97 - $154 per week ($5001 - $8000 per year)	❏
D.	$155 - $230 per week ($8001 - $12000 per year)	❏
E.	$231 - $308 per week ($12001 - $16000 per year)	❏
F.	$309 - $385 per week ($16001 - $20000 per year)	❏
G.	$386 - $481 per week ($20001 - $25000 per year)	❏
H.	$482 - $577 per week $25001 - $30000 per year)	❏
I.	$578 - $673 per week ($30001 - $35000)	❏
J.	$674 - $769 per week ($35000 - $40000)	❏
K.	$770 - $961 per week (40001 - $50000)	❏
L.	$962 - $1154 per week ($50001 - $60000)	❏
M.	$1155 - $1346 per week ($60001 - $70000)	❏
N.	>$1346 per week (>$70000 per year)	❏
O.	Don't Know	❏
P.	Refusal	❏

HOUSEHOLD RECORD FORM

O. What was the secondary carer's income before tax per week or per year in the financial year 1994-95?
- include family allowance and other benefits, child support, superannuation, wages and salaries, overtime, dividends, business income, interest.
- do not deduct tax, superannuation payments, health insurance payments.

Interviewer: Tick one appropriate category

Unprompted

A.	<$58 per week (<$3001 per year)	☐
B.	$58 - $96 per week ($3001 - $5000 per year)	☐
C.	$97 - $154 per week ($5001 - $8000 per year)	☐
D.	$155 - $230 per week ($8001 - $12000 per year)	☐
E.	$231 - $308 per week ($12001 - $16000 per year)	☐
F.	$309 - $385 per week ($16001 - $20000 per year)	☐
G.	$386 - $481 per week ($20001 - $25000 per year)	☐
H.	$482 - $577 per week $25001 - $30000 per year)	☐
I.	$578 - $673 per week ($30001 - $35000)	☐
J.	$674 - $769 per week ($35000 - $40000)	☐
K.	$770 - $961 per week (40001 - $50000)	☐
L.	$962 - $1154 per week ($50001 - $60000)	☐
M.	$1155 - $1346 per week ($60001 - $70000)	☐
N.	>$1346 per week (>$70000 per year)	☐
O.	Don't Know	☐
P.	Refusal	☐

HOUSEHOLD RECORD FORM

Appendix 4.6

Lung Function and Medication

Recording Sheets

File No		
Person		
Gender		
Group		

	day	month	year
DOB			
Date			

LUNG FUNCTION

PRE TREATMENT

	ABSOLUTE BEST/PRE	% PRED/NORM
FVC (litres)		
FEV$_1$ (litres)		
FEF $_{25-75\%}$ (litres/sec)		

MEDICATION	TYPE	DOSAGE
ENZYMES		
ANTIBIOTICS		
VITAMINS		
OTHER		

Appendix 4.7

Activity Recording Sheet

The following section is to help us estimate how much energy your child with CF uses up each day. This section is not asking what your child should do. It is asking what they currently do.

DOB

Date

day   month   year

Tell me what type of chest drainage or physiotherapy, exercise and sport your child with CF currently does. Do they do .... ?

TECHNIQUE TYPE	WHO DOES THIS? The Child, Mum, Dad etc	HOW OFTEN IS IT DONE WITH THE CHILD? times/day or times/week	FOR HOW LONG? minutes per session	AMOUNT OF SPUTUM PRODUCED 5ml=1tsp
Active Cycle of Breathing Techniques				
Forced Expiration Technique				
Positive Expiratory Pressure Mask				
Percussion (eg patting, tapping, clapping, thumping, tap-taps, tappies)				
Vibrations, Shaking				
Huffing				
Coughing				
Flutter Valve RP1				
Exercise Specify _____				
Sport Specify _____				

Other Specify _____ _____				
Other Specify _____ _____				

How old was your child when you were told he/she had cystic fibrosis?

months of age

In the past three months, has your child with CF

	Yes	No
started or changed oral antibiotics due to a lung infection?	☐	☐
started inhaled antibiotics due to a lung infection?	☐	☐
started IV antibiotics due to a lung infection?	☐	☐
been on oral steroids	☐	☐

CHEST DRAINAGE/ENERGY EXPENDITURE

Appendix 6.1

Revised Program Implementation

Implementing the Go and Grow with CF program

Six months before the introductory workshop:
- CF clinic staff were advised about the program
- introductory and concluding workshop dates during school holidays were selected. The dates coincided with CF clinic days for the convenience of patients who lived greater than 50 km from PMH. Several workshops were scheduled to ensure group size promoted interaction (ten to 15 families per workshop)
- workshop venues were reserved
- a list was prepared of children in the target group of two to 11 year olds
- a brief written explanation about the Go and Grow with CF program was posted to families and the dietitian encouraged families during CF Clinic appointments to participate in the three month program
- CF Clinic appointments were rescheduled to be on the same day as a workshop for patients who lived greater than 50 kilometres from PMH, and
- sponsorship was sought, from associations and pharmaceutical companies involved in the care of children with CF, for the home-based course materials and workshop expenses (approximately $10 per child).

Two months before the introductory workshop:
- volunteer dietitians and nutrition students were recruited to conduct the children's workshops, while the CF clinic dietitian would be conducting the parents' workshops
- care for children less than six years old during the workshops was arranged through the PMH Volunteer Service, and
- visits to the workshops by celebrities, such as national football player, were arranged through the PMH Public Relations Department.

Six weeks before the introductory workshop:
- an invitation to participate in the program was sent to families, and
- the home-based course folders for each patient, and material for the workshops, were prepared.

Three weeks before the introductory workshop:
- a list was prepared of the families who had indicated their intention to attend
- a letter was sent to carers to confirm workshop details, and
- CF clinic staff were informed about workshop and home-based course dates.

At the introductory workshop:
- the home-based course folders were presented to children or carers. A home-based course folder and written explanation about implementation were posted to families who did not attend the workshop.

During the home-based course:
- newsletters were prepared and sent fortnightly to complement the Go and Grow with CF weekly activities
- CF clinic staff encouraged families during appointments to continue participating in the program
- the dietitian phoned carers once per month to motivate families to complete the Go and Grow with CF activities, and to collect information for process evaluation, and
- families sent completed worksheets to the dietitian in pre-stamped addressed envelopes for review half-way through, and at the end of, the course.

Six weeks before the concluding workshop:
- an invitation to participate in the workshop was sent to families, and
- venues, celebrities, volunteer dietitians, nutrition students and child carers were confirmed.

Two weeks before the concluding workshop:
- a list of participants for the workshops was prepared
- a letter was sent to carers to confirm workshop details, and
- equipment for the workshops was arranged.

At the concluding workshop:
- achievement certificates were awarded to children and their families for participating in Go and Grow with CF. Families who did not attend a concluding workshop received their certificate at CF clinic appointments.

Appendix 6.2

Process Evaluation Data Recording Sheets

for the Revised Program

Since the beginning of this program / Since I last phoned:

Name	Date	What activity are you currently doing? (1-11)	What new thing have you learned from 'Go and Grow with CF'?	What new thing has your child learned from 'Go and Grow with CF'?	What have you enjoyed in 'Go and Grow with CF'?	What has your child enjoyed in 'Go and Grow with CF'?	What have you not liked in 'Go and Grow with CF'?	What has your child not liked in 'Go and Grow with CF'?

Since the beginning of this program / Since I last phoned:

Are you using the reward system every week? Y=yes N=no If yes, what rewards has your child chosen. If no, explore reasons why not.	Have you or your child had any nutrition or enzyme problems since starting the program? If yes, what? Encourage all parents to use the steps for problem solving sheets, if necessary.	If the family have identified a problem go through the steps for problem solving sheet with them. They may have filled one in already. Write any relevant comments in this box.	For parents of children 4 yo or more: Does your child swallow enzyme capsules whole? Y/N If no, discuss plans for learning to swallow capsules.	Have you put into action any of the information from the Supplement for Parents section pages 50-58? If yes: what? Encourage all parents to read it a couple of times.	What have you found difficult about the 'Go and Grow with CF' program?	What has your child found difficult about the program	What improvements do you suggest for the 'Go and Grow with CF' program? What could make it better?

Describe how you felt about the program before you started. 1.very uninterested 2.uninterested 3.indifferent 4.interested 5.very interested	What is your level of motivation to keep doing the program? 1.very unmotivated 2.unmotivated 3.indifferent 4.motivated 5.very motivated	If 1, 2 or 3 Why?	If 1, 2 or 3 What could improve your motivation?	What do you think is your child's level of motivation to keep doing the program? 1.very unmotivated 2.unmotivated 3.indifferent 4.motivated 5.very motivated	If 1, 2 or 3 Why?	If 1, 2 or 3 What could improve your child's motivation?	Other information

Name	Date	What activity are you currently doing? (1-11)	Since I last phoned... What new thing have you learned from 'Go and Grow with CF'?	What new thing has your child learned from 'Go and Grow with CF'?	What improvements do you suggest for the 'Go and Grow with CF' program? What could make it better?	Turn to page 3 What topics or activities were most beneficial for you in the new 'Go and Grow with CF' nutrition and enzyme program?	What topics or activities were most beneficial for your child in the new 'Go and Grow with CF' nutrition and enzyme program?	Our clinic policy is to continue matching enzymes to fat in food & drinks, BUT if you were given a choice which would you choose? 1= matching.....or 2= go back to taking enzymes as your child did before you learned matching 3= don't know Why?

What reinforcers / support from the dietitians / CF clinic staff worked best to keep you on track with the program? 1. Phone call 2. Newsletters 3. Conversations at clinic 4. Other, specify	What reinforcers / support from the dietitians / CF clinic staff did you not like? 1. Phone call 2. Newsletters 3. Conversations at clinic 4. Other, specify	Prior to the program, the goal you set for yourself was:	By the end of the program was this achieved? Y=yes N=no DK=Don't know If goal not achieved, ask, Do you know why?	Prior to the program, the goal you set for your child was:	By the end of the program was this achieved? Y=yes N=no DK=Don't know If goal not achieved, ask, Do you know why?	Would you recommend the 'Go and Grow with CF' program to other parents who have a child with CF? Y=yes N=no DK=Don't know Why?

Appendix 6.3

Faecal Fat Balance Study

Instructions and Sheets for

- Dietary recording

- Faecal collection

Note: For copyright reasons Appendix 6.3 has not been reproduced.

(Co-ordinator, ADT Project (Retrospective), Curtin University of Technology, 9.7.03)

Appendix 6.4

Faecal Fat Balance Study

Data Recording Sheets

File No.	Person No.	Gender 1 = M 2 = F	DOB	Name	Date	Height CM	Weight KG	Scale No.	Date Returned	Ranitidine Zantac	Miso-prostol Cytotec	Ome-prazole Losec	Cimet-idine Sigmet-adine	Cisipride Prepulsid	URSO Destolit	Bicar-bonate	Tube Feeds	Short Gut

Appendix 7.1

Faecal Fat Balance Study

Recommended Dietary Recording Sheet

Dietary and pancreatic enzyme record sheet (Fat balance)

SURNAME: _____   DAY: _____

FIRST NAME: _____   DATE: _____

DOB: _____   TYPE OF ENZYMES: _____

UMRN: _____

* Whenever possible, weigh food and
measure liquids using a metric cup

TIME	TYPE OF FOOD/DRINK	COOKING METHOD	* AMOUNT SERVED	* AMOUNT LEFT	* AMOUNT EATEN OR DRUNK	GRAMS OF FAT	NUMBER OF ENZYME CAPSULES

Appendix 8

Publications, Submissions, Presentations & Awards

Related to this Work

Publications

Stapleton D, Tunnecliffe L, McGuiness D, Sherriff J and Sly P. Development of a nutrition and behaviour intervention program: Go and Grow with CF. Aust J Nutr Diet 1998;55(3):130-137.

Stapleton D, Gurrin L, Zubrick S, Silburn S, Sherriff, J and Sly P. What do children with CF and their parents know about nutrition and enzymes? J Am Diet Assoc 2000;100(12):1494-1500.

Stapleton D, Go and Grow with CF (home-based course material for the nutrition and behaviour intervention program for children with CF) 1996, 1998.

Stapleton D, Go and Grow with CF: the implementation manual (for dietitans in CF centres) 1998.

Manuscripts submitted

Stapleton D, Gurrin L, Zubrick S, Silburn S, Sherriff, J and Sly P. The effect of Go and Grow with CF on nutrition and pancreatic enzyme knowledge of children with cystic fibrosis

Stapleton D, Kerr D, Gurrin L, Sherriff, J and Sly P. Height and weight fail to detect early signs of malnutrition in children with cystic fibrosis

Stapleton D, Mews C, Bulsara M, Sherriff, J and Sly P. Pancreatic enzyme replacement therapy: adherence to guidelines

Presentations

1996

Cystic Fibrosis Conference, Melbourne

Research and Advances Seminar, Princess Margaret Hospital, Perth

1997

16th Dietitians Association of Australia Conference, Hobart

American Thoracic Society Conference, San Francisco

Cystic Fibrosis and Nutrition Seminar, Royal Children's Hospital, Melbourne

Cystic Fibrosis and Nutrition Seminar, Great Ormond Street Hospital, London

Paediatric Nutrition Research in WA Seminar, Princess Margaret Hospital, Perth

Research and Advances Seminar, Princess Margaret Hospital, Perth

2nd Australian Cystic Fibrosis Conference, Perth.

1998

Australian Association for Cognitive and Behaviour Therapy Workshop, Perth

Dietitians Association of Australia WA branch and Australian Nutrition Foundation (WA Division) Joint Conference, Perth

17th Dietitians Association of Australia Conference, Sydney

22nd European CF Conference, Berlin, Germany

33rd Australian Psychology Society Conference, Melbourne

Australian Conference of Science and Medicine in Sport, Adelaide

Research and Advances Seminar, Princess Margaret Hospital, Perth

1999

3rd Australian Cystic Fibrosis Conference, Sydney

2000

Dietitians Association of Australia WA Branch, CPD Research Review Seminar, Curtin University of Technology, Perth

Awards

1996   Allied Health Travel Fund Prize, Princess Margaret Hospital

1997   The JA Mattinson Scholarship, Princess Margaret Hospital

1997   Curtin University Post-graduate Scholarship

1998   Janssen-Cilag Travelling Fellowship in Cystic Fibrosis

2000   Dietitians Association of Australia, WA Branch, Service Award for contributions to and expertise in nutrition in cystic fibrosis and co-convening the first 'National' Special Interest Group (CF) of the Association

Development of a nutrition and behaviour intervention program: Go and Grow with CF

Denise Stapleton, Lesley Tunnecliffe, Del McGuiness, Jill Sherriff and Peter Sly

Abstract The relationship between nutritional status and pulmonary function, acute infections and length of survival of patients with cystic fibrosis (CF) is well documented. Children with CF require 120% to 150% of normal energy requirements to achieve adequate weight gain and growth. The Go and Grow with CF nutrition and enzyme program was developed to combine dietary education with strategies for behavioural change. The program targets primary school-aged children with CF and their carers, but also includes information and strategies useful for carers of two- to five-year-olds. Six steps, based on social learning theory constructs, are used in the program to provide a framework for establishing new behaviours. Twenty-one children aged six to 11 years, and carers of 29 children aged two to 11 years, were enrolled in the program in 1996. Eighty-two per cent of children and/or their carers completed the course. Carers reported that they and their children learned about the correct use of enzymes, foods high in fat and salt, and the signs of malabsorption. Key features of the program were flexibility: learning in the home environment; and a structured, integrated system for reinforcing behaviour change. (Aust J Nutr Diet 1998;55:130–137).

Keywords: nutrition, pancreatic enzymes, education, program, behaviour, cystic fibrosis, children, pancreatic enzyme replacement therapy.

Introduction

Cystic fibrosis (CF) is the most common genetically-inherited disease in the Caucasian population. The disease affects the respiratory, gastrointestinal, pancreatic and reproductive systems, and causes elevated sweat secretions of electrolytes [1]. The problems of malnutrition and inadequate growth continue to exist despite advances in medical treatment, pancreatic enzyme preparations and dietary advice [2–6].

Factors likely to be responsible for causing energy deficits in patients with CF, and subsequent sub-optimal growth, include inadequate dietary intake, increased faecal losses, chronic respiratory infections and increased energy requirements [7]. The interaction between these factors becomes evident with worsening lung disease. Associated anorexia results in energy deficits. In turn, subsequent poor nutritional status contributes to progressive deterioration of lung function [7].

Energy-related nutrition recommendations suggest that patients with CF should consume 120% to 150% of normal daily energy requirements, based on age and gender [8,9]; obtain 40% of the energy requirement from fat [10]; and consume at least 100 g of fat per day, if they are of primary school age [11].

These recommendations are supported by studies demonstrating improved weight gain and lung function in patients with CF who were supplied with extra energy via supplemental tube feeding [12–15]. Numerous dietary assessments of children with CF indicate that they consume only 80% to 110% of normal energy require-

ments [5,16–20]. This suggests they are not able to meet elevated energy requirements without supplementation.

The inadequate oral intake of patients with CF may be the consequence of poor knowledge regarding their requirements. An assessment of CF patients aged ten to 16 years revealed a lack of knowledge about diet and enzyme use, despite patients and their immediate families receiving regular specialist dietetic advice [21]. Poor knowledge may be compounded by inappropriate parent and child behaviours at meal times [17,22–24]. Equipping parents with behaviour management strategies, in addition to educational information, may increase the opportunities for achieving optimum nutrition and growth in children with CF [25–27].

In addition to a high energy, high fat diet, children with CF and their parents must also learn how the child can achieve a high salt intake to compensate for elevated sweat losses of electrolytes [28,29]. Furthermore, up to 90% of patients have pancreatic insufficiency and require pancreatic enzyme replacement therapy (PERT) [30]. Recommendations regarding enzyme dosage, distribution, administration, timing and storage need to be learned and adhered to if maximum absorption is to be achieved.

Traditionally, in most Australian CF centres, families are counselled on the nutrition and pancreatic enzyme replacement needs of the child at diagnosis and, thereafter, at outpatient clinics or during hospital admissions. Information is presented in verbal and written form and is mostly directed to parents. The retention of information during these sessions may be limited by a number of factors. Initially, comprehension could be impaired by the emotional distress experienced at the time of diagnosis [31]. At clinics and during hospitalisation, the provision of too much information or fragments of information as problems arise may make it difficult for

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families to understand the relationships between concepts (32). This is likely if several members of the CF team provide numerous pieces of information to a family on the same day or the topic addressed is related to a crisis situation, such as inadequate growth. Without a comprehensive and accurate knowledge structure, families may be less able to apply information to address specific issues or problems (32).

Some centres in Australia conduct group education programs for children, separate from their parents. The literature supports the implementation of group-based programs to enhance a family's ability to implement strategies aimed to achieve adequate nutrition and minimise malabsorption (33,34). Group education programs, for children and parents, may help improve their understanding of the connections between concepts because information can be presented in a structured format. Participants can learn from, and be motivated by, one another and desirable behaviour and skills can be modelled (34-37).

Group programs have the additional benefit of being cost- and time-effective for health professionals (34). However, several weekly or fortnightly group sessions may not be convenient for parents and participation rates can decrease with time (35,38). When Bartholomew et al. (35) piloted the CF Family Education Project (FEP) only 23% of families attended at least one of four parts of the group-orientated program. Parents were pleased the education programs were available but could not devote the time to attend. Based on this feedback from parents, these authors wrote a program which was home-based (33).

Support for home-based programs is also found in a review of research on changing health-related behaviours within a family context. Perry et al. (38) considered the importance of parents in influencing children's behaviours when developing the Hearty Heart Home Team general nutrition program for children of primary school age. Those participating in the home-based program reported more diet-related behaviour change in line with the program objectives than those taking part in a similar school-based program.

Both Bartholomew et al. (35) and Perry et al. (38) used the social learning theory (SLT) as a basis for planning and developing comprehensive home-based education programs. SLT assumes people and their environments interact continuously (39,40). The key concepts of reciprocal determinism, behavioural capability, expectations, self-efficacy, observational learning and reinforcement address the psychological factors determining health behaviour and provide a basis for developing strategies to promote behaviour change (40).

The application of SLT constructs in the nutrition module of the CF FEP, for primary school aged children, appears to be limited to observational learning through modelling (33). The program does not provide comprehensive information on all aspects of nutrition and PERT, such as salt and vitamins (behavioural capability); or include other strategies, such as setting small goals and extrinsic reinforcers, which Perry et al. (38) suggest are important in a behaviour program for children (self-efficacy and reinforcement). Waters and Kennedy (41) outline three different types of rewards parents can use to reinforce desirable behaviours in children:

- verbal rewards, such as general praise statements, e.g. 'Well done!' or specific praise statements, e.g. 'Well done, you ate nearly all of your meal!' The use of specific praise helps children learn what is praise-worthy behaviour;

- physical rewards, such as a hug or a kiss. These can be coupled together with specific praise for extra emphasis on the desired behaviour; and,

- material rewards, such as charts, stickers, points and treats.

The inclusion of weekly rewards in a program, in addition to parental involvement, would maximise the likelihood of children with CF making major changes to food selections and adhering to PERT recommendations. External incentives (such as newsletters and regular contact with nutritionists) are also required to maintain parental involvement in a home-based program (38).

In 1996, the Go and Grow with CF education and behaviour change program was developed for children with CF and their carers (the adult most responsible for the child's CF nutrition needs and PERT). This program differs from the CF FEP (33) in the application of three SLT concepts. Go and Grow with CF provides:

- comprehensive information targeted for children of primary school age (behavioural capability);

- a system of small incremental steps to facilitate behaviour change (self-efficacy); and,

- a greater range and more structured approach to rewards (reinforcement).

The aim of this paper is to describe the development of Go and Grow with CF, discuss features of the program and report findings of the process evaluation.

Methods

Program development

Focus groups

Focus groups were conducted to assess the need for a change in the approach to CF nutrition and PERT education at Princess Margaret Hospital for Children (PMH). Carers of children scheduled to attend two CF clinic sessions were invited to participate in a nutrition-related focus group discussion. The focus groups lasted for one hour and were conducted before the CF clinic sessions. Each focus group involved approximately eight carers of two- to 11-year-olds, who met separately from approximately five children aged seven to 11 years. Carers and children were interviewed by health professionals not involved in the CF clinic.

The focus group discussions identified a number of issues.

- Carers were interested in learning more about nutrition and enzymes, and agreed a new approach to nutrition education would be worthwhile.

- Most carers enjoyed meeting with other families to discuss aspects of caring for a child with CF.

- Carers preferred teaching sessions to be held on the same day as clinic sessions to minimise the number of trips to the hospital, particularly for rural families.

- Carers often- to 12-year-old children wanted children involved in an education program from a young age.

- Carers thought most interest in a new nutrition program would come from families whose child had been diagnosed with CF in the last four years.

- Carers identified child-care facilities for siblings as an important factor in promoting participation in group sessions.

The focus groups and review of the literature provided sufficient support for a new approach to nutrition and PERT education at PMH.

The model

The Go and Grow with CF program was developed by a dietitian and a psychologist to facilitate families in translating nutrition and PERT knowledge into behaviour and action. The program was designed for children of primary school age (six to 11 years), and carers of two- to 11-year-olds, and combined group sessions with home-based learning.

The focus of Go and Grow with CF was learning in the home environment, with particular emphasis on the SLT concepts of behavioural capability, self-efficacy and reinforcement [39,40]. Children and carers were guided through six steps for each of seven nutrition topics to help establish new behaviours (see Figure 1).

The concept of behavioural capability was addressed by providing comprehensive nutrition and PERT information in the home-based course. This information would equip families with the knowledge and skills necessary to be capable of achieving desirable behaviours.

Figure 1. The 'Go and Grow with CF' program structure

Process	Social learning theory constructs
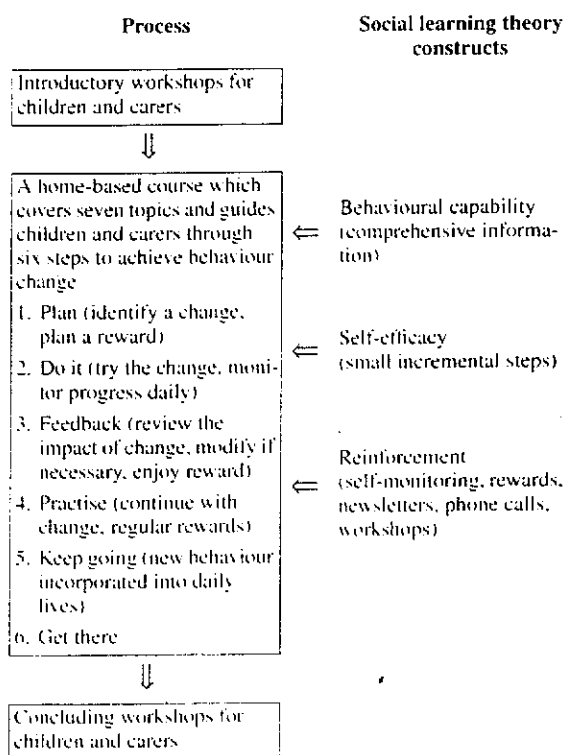
Participants were guided to set small, incremental goals during the home-based course to help increase their self-efficacy. For example, at the beginning of the energy and fat topic, children were guided to plan a high energy, high fat breakfast every day for a week. The following week the focus was high energy, high fat morning snacks. The children were challenged to consume these more often than they had consumed their planned breakfasts the week before. A third week was spent on high energy, high fat supper snacks. The purpose of planning daily meals or snacks, and challenging children to consume these more often each week, was to help them reach the long-term goal of a high energy, high fat intake every day.

Social reinforcement was provided by carers becoming involved in the children's program. Weekly rewards, for participation with daily plans, provided external reinforcement. Carers and children were asked to prepare the reward list together to ensure the activities and items were valued by the child [41,42].

Another source of reinforcement in Go and Grow with CF was the task of recording adherence to daily plans. This activity of self-monitoring provided children with immediate feedback about their ability to change, and was a basis for the dietitian to provide external reinforcement when completed worksheets were reviewed once a month. Additional external reinforcement for families came in the form of fortnightly newsletters and monthly telephone contact between the carer and the dietitian during the home-based course.

The program

The Go and Grow with CF program is illustrated in Figure 1. The program began with separate, concurrent introductory workshops for children of primary school age and for carers of two- to 11-year-olds to introduce nutrition and PERT learning objectives; explain the home-based course; and motivate families to participate and complete the program.

The workshops were followed by the home-based course, which consisted of printed materials and ten weekly activities for children and carers to complete together. The seven topics covered in the home-based course were: enzymes, energy and fat, malabsorption, vitamins and minerals, growth, snacks, and salt. Finally, separate, concurrent concluding workshops for children and carers were held to review their learning and celebrate participation and progress throughout the program.

The introductory workshops were conducted during a school holiday, the home-based course was completed during the school term, and the concluding workshops were held during the following school holiday. The workshops for carers and children were held in an informal setting at the hospital and were conducted by the CF clinic dietitian and other dietitians who participated on a voluntary basis. The written material for the home-based course was presented to families at the end of the children's introductory workshop. Families who did not attend the introductory workshop received the home-based course by post and implementation advice by telephone.

The home-based course was designed to meet all the nutrition, enzyme and behaviour objectives of the program. The child and carer met once each week to:

- review their participation and/or progress with the previous week's topic;

- discuss strategies to improve or continue practising their new skill;

- decide when to enjoy their reward for participation and/or progress during the previous week;

- read information and complete the activities for the next topic;

- plan a reward for participation and/or progress for the following week; and,

- plan what they would do each day of the coming week relevant to the topic.

During the week, the family recorded whether or not they achieved the planned changes each day. The home-based course took families approximately one hour per week to complete.

The participants

A clinical trial was conducted to assess the impact of Go and Grow with CF. Exclusion criteria included pancreatic sufficiency, severe liver disease requiring drug therapy, short gut syndrome, enteral tube feeding and marked language delay. A total of 70 children were eligible, but 16% did not wish to participate or attended the CF clinic irregularly. Of the 59 children aged two to 11 years who were enrolled in the clinical trial, 30 were randomly selected to receive the Go and Grow with CF program. One carer chose not to participate due to time constraints. Program participants included 21 children aged six to 11 years and 27 carers of 29 children aged two to 11 years.

Process evaluation

Participation rates were calculated as a proportion of those who agreed to take part in the program. Carers' program goals for themselves and for their children were documented during the introductory workshop, or after implementation advice was provided by telephone. At the completion of Go and Grow with CF, carers indicated whether or not these program goals were achieved. During telephone contact with the dietitian at weeks 4, 7 and 10 of the home-based course, carers self-reported what they, and what they thought their child, were learning, enjoying and disliking. Carers were also asked to indicate the effect of the program on their family; what helped them to continue participating in the home-based course; and whether they would recommend Go and Grow with CF to other families who have a child with CF. As the questions were open-ended, percentages are not reported for response categories. Assigning percentages would suggest carers were given an opportunity to comment on each category in turn (43). A record of dietetic time spent conducting all aspects of the program was maintained over three months.

Ethics approval for the program and clinical trial was obtained from the Princess Margaret Hospital Research and Ethics Sub-Committee and the Curtin University of Technology Human Research Ethics Committee.

Results

Participation

Only 57% of children and 50% of carers attended both workshops. Ten per cent of children and 30% of carers did not attend either workshop. Carers' reasons for not attending the workshops were related to not wanting to attend extra hospital activities, having work commitments or family illness. Seventy-three per cent of children and/or their carers completed the home-based course on schedule, i.e. before the concluding workshop. Family illness, such as chickenpox, and being busy, temporarily interrupted 10% of families. Three weeks after the concluding workshop a total of 82% of children and/or their carers had completed the home-based course (22 carers, 17 children). Reasons given by carers who did not complete the home-based course were related to being too busy or having extra demands on their time from newly-born or disabled siblings. All these carers expressed the desire to participate in the program at a later stage.

Process evaluation

Results of the process evaluation are reported for the group of 82% of children and/or their carers who completed the home-based course.

Goals

The goals carers set for themselves and for their children were related to nutrition and/or pancreatic enzyme requirements of a child with CF. The most common goals carers set for themselves were:

- to learn more about food, nutrition, enzymes and malabsorption;

- to help their child gain weight, eat faster, keep healthy and avoid bowel blockages;

- to manage their child's diet better, to be more adaptable with their child's food and to be one step ahead of their child in the area of food;

- to learn how to deal with nutrition and enzyme problems; and,

- to learn how to maintain their child's interest in treatment, to help their child understand CF-related health needs and to be satisfied that their child has been informed about his or her CF needs.

The most common goals carers set for their children were:

- to learn more about CF nutrition needs, high fat foods, enzymes and their importance, and malabsorption;

- to be more responsible about nutrition and enzyme needs; and,

- to remember to take enzymes.

Ninety-six per cent of carers reported achieving the goal they set for themselves. Eighty-five per cent of carers reported the goal they set for their children was achieved.

Learning

Ninety-one per cent of carers reported learning at least one aspect from a range of topics, including pancreatic enzymes, foods high in fat and salt, increasing energy intake and malabsorption. Ninety-three per cent of carers perceived their children were learning from the program.

The nutrition and enzyme concepts that carers reported they learned were: the correct use of enzymes and that enzymes are active for 30 minutes; what foods are high in fat and how to increase energy intake; the need for salt and what foods are high in salt; the signs of malabsorption; and, topics reinforced and placed into perspective.

The nutrition and enzyme concepts that carers reported their children learned were: the correct use of enzymes and the importance of taking enzymes; the signs of malabsorption; the need for food high in fat; and, the need for salt and what foods are high in salt.

Enjoyment

Ninety-one per cent of carers enjoyed at least one aspect about the course, such as helping their child learn, learning themselves, or gaining confidence from a better understanding of nutrition and PERT. Aspects of the home-based course that carers reported children most enjoyed were the rewards for participation each week, monitoring every day (ticking boxes, drawing smiling faces) and spending time with their carer. Rewards which were most successful in maintaining participation in Go and Grow with CF varied. Responses were equally spread over activities (having friends to sleep over, board games, computer time), objects (stickers, toys), money, and food treats (fast food, lollies). Some children were not interested in material rewards. These children enjoyed daily monitoring of their progress on the worksheets or spending time with their carer doing the program.

Dislikes

During the home-based course, carers reported they disliked being too busy to easily fit the program into the week, daily recording or not having planned food available. Thirty-six per cent of carers could not identify aspects they disliked about the program. Some carers thought children disliked their play-time being interrupted to do the worksheets or daily recording.

Effect on the family

Fifty per cent of carers reported that 'Go and Grow with CF' had a positive effect on the family, such as sibling interest in CF nutrition and PERT needs. Nine per cent of carers reported the program had a negative effect on the family, such as a sibling requesting high energy, high salt food at the beginning of the home-based course.

Motivation

Telephone calls and fortnightly newsletters from the dietitian motivated carers (95% and 62% respectively) to continue participating in the home-based course.

Recommendation to others

All carers who participated in the program indicated they would recommend Go and Grow with CF to other families with a child who had CF. Reasons for recommending the program included benefits related to both knowledge gain and behavioural change. Reported benefits that related to knowledge gain were:

- much was learned from the program that was not known before;

- the program was helpful with young children and especially early in diagnosis;

- children learnt about nutrition and enzyme needs;

- it was a good revision process;

- the program was more informative than previous information;

- it was set out in plain language; and,

- there was increased awareness of what children are eating and what needs to be added.

Reasons related to behavioural change were that the program:

- improved habits and involved the child;

- helped parents make the effort to provide appropriate food; and,

- helped parents work out nutrition and enzyme problems.

Dietetic time

An average of two and a half hours per family was spent by the dietitian conducting the Go and Grow with CF program over three months. The time was divided between workshops, telephone contact, newsletters and providing feedback on completed worksheets.

Discussion

This process evaluation of the Go and Grow with CF program indicates a high rate of participant satisfaction and perceived learning. Workshops provided a group learning experience and enabled the dietitian to orientate carers to concepts in the home-based course. The low workshop participation rates did not appear to limit the completion rate of the home-based course. Half of the carers who did not attend either workshop did complete the home-based course. This suggests factors other than the workshops provided sufficient motivation during the program. These factors are likely to have been the desire of the carers to learn, monthly telephone contact with the dietitian, and fortnightly newsletters.

Involvement in the program enabled participants to meet almost all the carers' goals for themselves and their children. Carers recognised some of the goals not achieved were inappropriate for the young age of the child (e.g. wanting a five-year-old to always remember to take enzymes).

A majority of carers perceived they and their children had learned new concepts from Go and Grow with CF and carers appeared to enjoy the learning process. Children enjoyed behaviour change reinforcers, such as rewards, monitoring and carer involvement.

Some carers reported they or their children disliked daily monitoring or being too busy to easily fit the program in each week. Newsletters and telephone contact with the dietitian appear to have helped families to be motivated to incorporate the program into busy schedules.

Go and Grow with CF was rated highly by participants as all carers indicated they would recommend the program to other families who have a child with CF.

The generalisability of the results obtained in this study are limited by the potential bias caused by the likeli-

hood of carers enrolled in the clinical trial having a particular interest in nutrition-related aspects of CF, and by the program developer conducting the process evaluation.

Participation and completion rates of the home-based course may have been lower if the target group included irregular clinic attendees (who attended less than once per year) and carers who disliked hospital-related activities. Such families were not involved in this study as they declined the offer to be enrolled in the clinical trial. Carers may have been less inclined to report positive responses about Go and Grow with CF if they reported their perceptions to someone other than the person who developed the program. Future assessment of growth and behaviours will provide more objective measurements of the success of the program.

Flexibility was a key feature of Go and Grow with CF. This flexibility accommodated different lifestyles, learning styles, medical needs and minimised competition between the program and a busy family life. The home-based course allowed families to determine the best time for them to complete the worksheets and probably contributed to the high completion rate.

Sickness, unrelated to CF, forced some families to interrupt the home-based course. The flexible nature of the program allowed them to recommence a few weeks later, without having missed any information. The families considered the illness as a temporary interruption and completed the home-based course soon after the concluding workshop. This is an important aspect of the Go and Grow with CF model considering the increased likelihood of illness with a chronic disease.

At the conclusion of the program it became evident that the flexibility of Go and Grow with CF provided an opportunity for other family members to be educated. A high fat, high salt diet is unsuitable for people who do not have CF. Participation in the program may help siblings understand that the dietary needs of the child with CF are part of their therapeutic treatment. Siblings may be more willing to accept diet-related inequalities in the home when they are better informed. Weekly meetings with several members of the family, to complete the home-based course, would also provide an opportunity to enhance the learning of children who prefer group-based learning over individual learning.

Finally, it became evident while reviewing the home-based course that its flexibility could allow the program to be adapted for the small percentage of children with CF who have pancreatic sufficiency. The topics which explore energy and fat, vitamins and minerals, salt and growth are relevant to all children with CF. Children who have pancreatic sufficiency could complete these topics but omit or only read the worksheets on enzymes and malabsorption.

Learning in the home environment was another key feature of Go and Grow with CF as health behaviour changes of children of primary school age appear to be maximised by involving their carers (38). Carers reported they and their children learned how to achieve a high energy, high salt diet and minimise malabsorption by participating in the program. It is paramount that children with CF and their carers understand and apply these fundamental concepts in order to achieve optimum growth and health. Previous learning and retention of information

may have been affected by the emotional distress carers experienced at diagnosis (31); the numerous distractions and tasks undertaken by children and carers during outpatient clinics and hospital admissions; and standard nutrition information indicating what to do, but not providing families with a system to enhance their self-efficacy and subsequent behaviour change.

Possible reasons why carers and children may have understood concepts more clearly through participation in Go and Grow with CF include the following:

* the Go and Grow with CF program provided comprehensive nutrition and PERT information which assisted carers in formulating a structured conceptual framework;

* the home-based course maximised involvement of carers as role models, teachers and reinforcers of behavioural change;

* the information and activities in Go and Grow with CF were in a format appropriate for children of primary school age; and,

* the inclusion of group sessions provided an opportunity for participants to learn from, and be motivated by, one another to complete the home-based course.

The use of a reward system to reinforce behaviour change was also a key feature of Go and Grow with CF. The introductory workshop provided an avenue for explaining the reward system as this was a new concept for many carers or one not fully understood by others. Some carers needed extra assistance to help identify rewards valued by the child and to ensure the system was initially used only for behaviours related to the program. The provision of detailed information and steps to follow to establish the reward system in the revised version of the program will ensure carers, including those who do not attend the introductory workshop, are well equipped to implement this approach, which may be new to them.

Successful implementation of Go and Grow with CF was dependent on the motivation of the dietitian to conduct the program, and adequate time allocation over three months. At PMH, the CF clinic dietetic time available for each patient averages two hours per year. The time required to conduct Go and Grow with CF was approximately two and a half hours per child over three months. The extra time invested in conducting the program could be recouped over subsequent years. Carers reported that Go and Grow with CF helped them develop skills to effectively deal with fundamental nutrition and pancreatic enzyme issues and problems. Although this is yet to be demonstrated, the program is likely to reduce the demands on dietetic staff of ad hoc problem solving, resulting in better use of expertise.

An additional saving on the dietitian's time could be achieved by conducting the program for all families in the target audience once every four years, rather than every year with a portion of the target group. Time spent preparing for the workshops, newsletters and evaluation would be spread across a larger number of participants. Clinics with a large population would need to conduct several introductory and concluding workshops (with ten to 15 families per workshop) to ensure group size promoted interaction.

The process evaluation and recent advances in CF nutrition and PERT recommendations indicated the program needed the following modifications:

- the addition of comprehensive information for carers about using a reward system;

- guidelines for using the program with children who have pancreatic sufficiency;

- guidelines for successfully involving other family members;

- the addition of the 1997 Australian PERT guidelines (44);

- incorporation of a target for primary school-aged children with CF to consume 100 g of fat per day; and,

- further process evaluation, including pre- and post-measurements of dietetic time spent in ad hoc problem solving.

Conclusion

Go and Grow with CF used social learning theory methods to assist primary school-aged children with CF and carers of two- to 11-year-olds in the process of translating nutrition and enzyme knowledge into behaviour and action. The program combined workshops with home-based learning to maximise the effect and appeal of the program to families. Key features of Go and Grow with CF were flexibility; learning in the home environment; and a structured, integrated system of reinforcing behaviour change.

Based on carers' high level of satisfaction with the program, and reports of learning fundamental nutrition and pancreatic enzyme concepts, Go and Grow with CF has been incorporated into the dietetic service provided to children attending the PMH CF clinic. The program has since been revised to include findings from the process evaluation and the 1997 Australian PERT Guidelines.

The strategies used in Go and Grow with CF have the potential to be successfully applied to various programs for families living with other chronic conditions; such as for children and adults with metabolic disorders or renal disease. The focus of the program could be nutrition, medication or physiotherapy practices. The Go and Grow with CF model could also be used to develop health promotion programs for families in the general community.

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Nutrition education

Online nutrition education in an Aboriginal community: the example of Wirura Nyinanyi

Alan Barnes

Successful nutrition education in Aboriginal communities requires strong partnerships between the educators and the community. The Anangu Teacher Education Program (AnTEP) at the University of South Australia has had a long involvement with traditionally-oriented Aboriginal communities south of Alice Springs, mainly in the Anangu Pitjantjatjara Lands and at Yalata. Its aim is to develop teachers who can bring their cultural experience to presenting effective teaching programs in Anangu schools (1) and as such can have a considerable impact on nutrition education in the region.

In 1997 the Aboriginal Research Institute at the University of South Australia received an Australian Research Council grant to place educational and research infrastructure into AnTEP sites in the Anangu Pitjantjatjara lands. A major task of the ongoing project is to bring high bandwidth digital connectivity to educational, health and research interests who have programs and projects in partnership with the Anangu Pitjantjatjara Council. One such partnership is being developed with CSIRO Human Nutrition and another with the South Australia Centre for Rural and Remote Health. The rationale is to allow distance research and teaching and to provide Anangu students with similar access to Internet services that are available on university campuses. Final deployment is expected in late 1998.

Within the framework of the infrastructure project, AnTEP and the Aboriginal Research Institute have developed an online subject called Wirura Nyinanyi (2), an Anangu concept that can be roughly translated into English as 'health and wellbeing'. The subject attempts to embody much of what has been learned by AnTEP staff about Anangu learning styles and practical aspects of distance learning. A substantial part of the subject is focused on nutrition. The subject will be run in Semester 2 1998.

As the screen shot indicates (Figure 1), the content of Wirura Nyinanyi is presented as standard web pages including navigation buttons, pictures, sound and movies.

The general design philosophy of the module has been to give expression to Anangu voices on nutrition while developing the English articulation of nutrition concepts. Assessment for the subject is in English. The online content includes clips from videos used by Nganampa health, pictures of the community store, Anangu songs and comments in Pitjantjatjara language (3-5). Comments by current and former students have been particularly useful in contextualising issues. Dual language oral translations of important text materials are included. Interaction is achieved through short exercises and through English writing that is relayed to a database in Adelaide. The stu-

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The effect of 'Go and Grow with CF' on nutrition and pancreatic enzyme knowledge of children with cystic fibrosis

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Abstract A randomised clinical trial was conducted to assess the impact of the 'Go and Grow with CF' intervention program on nutrition and pancreatic enzyme knowledge and self-management skills. Forty-two children with cystic fibrosis aged six to 11 years and 55 carers of two- to 11-year-old children participated in the program. Separate but similar validated questionnaires were used for face-to-face interviews with children with cystic fibrosis and for telephone interviews with carers on three occasions. The questionnaires were designed to assess nutrition and pancreatic enzyme knowledge and self-management skills. The improvement in knowledge of the children's intervention group was significant immediately post-intervention ($P = 0.001$) but not at follow-up 12 months later. Children's knowledge for both the intervention and control groups was associated with their appropriate self-management score immediately ($P = 0.02$ and $P = 0.03$, respectively) and 12 months post-intervention ($P = 0.01$ and $P = 0.00$, respectively). The home-based 'Go and Grow with CF' program was found to be effective in increasing children's knowledge in the short-term. The absence of a long-term effect of the program on nutrition and pancreatic enzyme knowledge and behaviours may be due to the need for regular ongoing education and counselling. (**Aust J Nutr Diet 2001;58:164–168**)

Key words: nutrition knowledge, questionnaire, cystic fibrosis, behaviour, intervention

Introduction

Cystic fibrosis (CF) is a common, severe autosomal recessive disorder affecting many body systems [1]. The nutritional problems and consequences of the disease are multifactorial. Lung function, anorexia, pancreatic insufficiency and intestinal and biliary complications contribute to increased energy needs, inadequate intake and excessive nutrient losses [2]. Despite recent advances in the treatment of CF, poor growth and malnutrition continue to be significant problems and affect the length of survival of patients [3–9].

Individuals with CF need to consume a high energy, high fat, high salt diet and up to 90% of patients also require pancreatic enzyme replacement therapy (PERT) [10,11]. An adequate level of knowledge is needed by children and their parents in order to be capable of making appropriate food choices and to optimise pancreatic enzyme administration. Children's knowledge of nutrition and PERT is often poor despite the provision of specialist dietetic advice to families [12,13]. Knowledge and subsequent adherence to treatments for CF could possibly be improved by the provision of periodic, structured, age-appropriate education programs [14].

A nutrition education and behaviour intervention program, 'Go and Grow with CF', was developed in 1996 with the specific aim of facilitating primary school-aged children and carers of two- to 11-year-olds in the process of translating nutrition and enzyme knowledge into behaviour and action [15]. A carer was defined as the adult most responsible for the child's nutrition and PERT needs. Specific objectives of the intervention program were:

- to increase the nutrition and PERT knowledge and self-management skills of children with CF and their carers; and,
- to increase the number and range of strategies used by carers to cope with food and PERT-related behaviour problems.

A clinical trial was established to assess the impact of the 'Go and Grow with CF' intervention program on knowledge, behaviour, dietary intake and nutritional status. The purpose of this paper is to report on the nutrition and enzyme knowledge and self-management results obtained from the clinical trial.

Methods

Study participants

Children eligible for the clinical trial were those attending the Princess Margaret Hospital CF clinic who were aged between two and 11 years at the commencement of baseline data collection and who had pancreatic insufficiency. Exclusion criteria for the clinical trial were pancreatic sufficiency, liver disease requiring drug therapy, short-gut syndrome, enteral tube feeding and marked language delay in children aged six to 11 years.

Of the 88 children in the specified age group attending the CF clinic, 70 were eligible for the clinical trial. Of these, 11 were not enrolled in the study because they did

stage of development and disease status. The 'Go and Grow with CF' home-based course material has the potential to be used as both a preventative program and to help identify strategies for solving nutrition and PERT-related problems if they arise.

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