Mount Saint Vincent University
Department of Applied Human Nutrition

Body esteem, eating attitudes, and adherence in adolescents with cystic fibrosis from Atlantic Canada.

by
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• Jesus: who loves me no matter what and has helped me to get through all 190 pages of this document!

DEDICATION

This thesis is dedicated to the fight against cystic fibrosis.

To those who have fought and lost.

To those who continue to fight.

To those who are too tired to fight anymore.

To those who fight on our behalf.

To the hopes of a day when CF will be no more.

65 roses of yellow and red,
Made her so tired she had to stay in bed.
I looked all around but I couldn't find,
The 65 roses were all in my mind.

I've grown up, and I see,
that the only one thinking of roses was me.
And the reason the 65 roses came,
was because I was too young to understand the name.

65 roses, Cystic Fibrosis
Made it so tired she had to stay in bed.
65 roses, Cystic Fibrosis
I wish she had roses instead.

65 roses, Cystic Fibrosis
I wish she could come out to play.
Life one supposes is no bed of roses
But I wish she had roses instead.

-The Wolverines, 65 Roses

BODY ESTEEM, EATING ATTITUDES, AND ADHERENCE IN ADOLESCENTS WITH CYSTIC FIBROSIS FROM ATLANTIC CANADA.

ABSTRACT

A study was conducted to investigate the influence of body esteem, nutritional status, nutritional intake, and eating attitudes on adherence to dietary therapy and enzymes in adolescents and young adults with cystic fibrosis. Nine adolescents aged 12.5 to 18 years from the CF clinics at the IWK (Halifax, NS) and the Janeway (St. John's, NL) participated in the study. The participants completed the following surveys/tools: a Three-Day Food Diary, The Eating Attitudes Test-26 (EAT-26), The Body Esteem Scale for Adolescents and Adults (BES), and The Adherence Survey.

Nutritional analysis determined that study participants had an average caloric intake of 2174 Cal. Only one participant met the adjusted Estimated Energy Requirement (EER + 20%), and was classified as being adherent to dietary therapy. The average Body Mass Index (BMI_p) was at the 52^{nd} percentile, and the average percent Ideal Body Weight (%IBW) was 100. None of the participants were classified as stunted, according to their height-for-age, but one was classified as wasted, according to his/her BMI_p value.

BES scores determined that participants have a relatively positive body esteem, with a mean score of 52.67, 57.25% of the highest possible score. Eating attitudes were also positive, for the most part. However, EAT-26 scores did reveal that one participant was at risk of developing an eating disorder, having a score over 20.

In the study, adherence was defined as taking the same dose, or more, of a treatment as prescribed by a doctor and/or dietitian. Adherence to vitamins and nutritional supplements, when prescribed, was extremely poor, at 0%. Adherence to enzymes taken with meals and snacks was similar to rates in the literature, with 56% being adherent. Common reasons cited for non-adherence included forgetfulness, an absence of positive results, time and effort consuming, and a dislike of taking the treatment in front of others.

Due to the small sample size, two-tailed independent sample t-tests revealed no statistical significance between adherence, body esteem, eating attitudes, caloric intake, BMI_p, and %IBW. Therefore, results of the study indicate that in the population of all adolescents with CF, based on the results of this study, one cannot predict differences in BES, EAT-26, EER, BMI_p, or %IBW based on the individuals's adherence category.

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Section 1: Introduction

1.1. Overview

Canadian adolescents, particularly females, frequently display disordered eating behaviors (Canadian Paediatric Society [CPS], 2004). Popular media often portray the ideal body as being thin and near anorexic (CPS, 2004). Being exposed to these distorted messages places teenagers and young adults at risk for unhealthy eating behaviors. Dieting can result in nutritional deficiencies; particularly in iron and calcium, which can lead to a decrease in growth (Dietz & Hartung,1985), menstrual irregularities, including amenorrhea (Selzer, Caust, Hibbert, Bowes, & Patton, 1996), and even osteopenia and osteoporosis (Turner et al., 2001).Various risk factors for unhealthy eating behaviors include: high parental criticism, low self-esteem, vegetarianism in females, psychiatric disorders, and chronic illness resulting in body dissatisfaction (CPS, 2004).

The most common chronic fatal genetic disease in Caucasians is cystic fibrosis (CF). This disease attacks the respiratory, digestive, and reproductive systems, and occurs in approximately one in 3600 Canadians (Canadian Cystic Fibrosis Foundation [CCFF], 2003). Nutrition is very important to the management of CF since pancreatic insufficiency occurs at a rate of approximately 85-95% (Sinaasappel et al., 2002; Creveling, Light, Gardner, & Green, 1997; Bronstein et al., as cited in Orenstein, Winnie, & Altman, 2002; Wolfe, Tremlett, White, & Littlewood, 2001). The resulting inadequate production of enzymes, leads to fat and protein malabsorption and maldigestion (Bronstein et al., as cited in Orenstein et al., 2002; Pencharz & Durie, 1993).

Malnutrition resulting from such malabsorption and maldigestion tends to result in a low Body Mass Index (BMI). Having a positive nutritional status can help improve overall health and

lung function (Murphy, Wooton, Bond, & Jackson, 1991). To compensate for maldigestion and malabsorption, pancreatic enzyme supplements are taken with food and an optimal dietary intake is high in calories and fat. However, with media messages supporting a preference for slimness, non-adherence to pancreatic enzyme supplements, nutritional supplements, and/or diet recommendations may occur in order to achieve, or maintain, CF-induced slimness.

Minimal research exists exploring the relationship between how one feels about their body and adherence to diet recommendations, nutritional supplements, and enzyme replacement therapy in teenagers with CF. Increased research may help clinicians identify and address these issues, leading to improved nutritional status that allows those with CF to live longer, healthier lives.

1.2. Research Statement

This research study will investigate how body esteem, eating attitudes, nutritional status, and nutritional intake affects adherence to diet, vitamins, nutritional supplements, and pancreatic enzyme replacement therapy in adolescents with CF.

1.3. Research Objective

To assess relationships involving the following variables in teenagers aged 13 to 20, inclusive, with cystic fibrosis:

- Body esteem;
- Eating attitudes;
- Adherence;
- Nutritional intake; and

- Nutritional status.

Section 2: Abbreviations and Definitions

2.1. Abbreviations

AS	The Adherence Survey
BES	Body Esteem Scale
BMD	Bone Mineral Density
BMI/BMI_p	Body Mass Index/Body Mass Index Percentile
Cal	Calorie
CCFF	Canadian Cystic Fibrosis Foundation
CDC	Centre for Disease Control
CF	Cystic Fibrosis
CFPC	College of Family Physicians of Canada
CFTR	Cystic Fibrosis Transmembrane Conductance Regulator
CHNA	Community Health Nurses Association of Canada
CPS	Canadian Paediatric Society
DC	Dietitians of Canada
DRI	Dietary Reference Intake
EAT/EAT-26	Eating Attitudes Test/Eating Attitudes Test-26
EER	Estimated Energy Requirement
EFA	Essential Fatty Acid
FEV_1	Forced Expiratory Volume in 1 Second
FFM	Fat Free Mass
g	Gram
HBM	Health Belief Model
HFA	Height-for-Age
ht	Height
IBW/% IBW	Ideal Body Weight/ Percent Ideal Body Weight
IU	International Units
kg	Kilogram
L	Litre
mmol	Millimole
mg	Miligram
mRNA	Messenger Ribonucleic Acid
PAC	Physical Activity Coefficient
PEP	Positive Expiratory Pressure
PERT	Pancreatic Enzyme Replacement Therapy
PUFA	Polyunsaturated Fatty Acid
REE	Resting Energy Expenditure
TEE	Total Energy Expenditure
TPB	Theory of Planned Behavior
TPN	Total Parenteral Nutrition
VC	Vital Capacity
WFA	Weight-for-Age
WHO	World Health Organization
wt	Weight

2.2. Definitions

Apical Cells: cells located on part of the superior aspect of a body, organ, or part, or the pointed extremity of a conical structure such as the heart or lung (Merck Source, 2006).

Chorionic Villus Sampling: prenatal testing procedure offered in certain pregnancies for determination of the chromosomal profile of the fetus. The chorionic villi are part of the placenta. CVS involves removing a small sample of the placenta with a thin needle which is inserted through a woman's abdomen or with a catheter which is inserted through the vagina and cervix (Chromosomal Mosaicism, 2003).

Distal Intestinal Obstruction Syndrome: involves blockage of the intestines by thickened stool and occurs in individuals with cystic fibrosis. DIOS was previously known as meconium ileus equivalent (Wikipedia, 2006).

Epithelial Cells: cells covering the internal and external surfaces of the body, including the lining of vessels and other small cavities. The epithelium consists of cells joined by small amounts of cementing substances (Merck Source, 2006).

Forced Expiratory Volume in One Second: the volume exhaled during the first second of a forced expiratory maneuver started from the level of total lung capacity (Quajner et al., n.d.).

Meconium Ileus: Obstruction of the intestine (ileus) due to overly thick meconium, the dark

sticky material that is normally present in the intestine at birth and, after trypsin and other enzymes from the pancreas have acted on it, is normally passed in the feces after birth (Medicine.net, 2006).

Vital Capacity: the volume change of the lung between a full inspiration and a maximal expiration (Quajner et al., n.d.).

Section 3: Literature Review

3.1. Cystic Fibrosis

3.1.1. Overview.

Evidence of cystic fibrosis first emerged in the 1930's when Dorothy Anderson described 49 autopsies as having "fibrocystic disease of the pancreas" (Littlewood, 2002). Vitamin A was thought to be involved in the etiology of CF, since the changes found in epithelial cells were similar to those associated with vitamin A deficiency (Littlewood, 2002). In the early 1940's it became apparent that there was a genetic basis to the condition, which was then known as mucoviscidosis (Littlewood, 2002). Vitamin A deficiency was still hypothesized to be an important factor in the development of the disease, believing that it was needed for the full expression of the genetic defect (Littlewood, 2002). It wasn't until later that decade that the role of vitamin A was questioned, and an abnormal epithelial secretion was then thought to be the cause of the disorder (Littlewood, 2002).

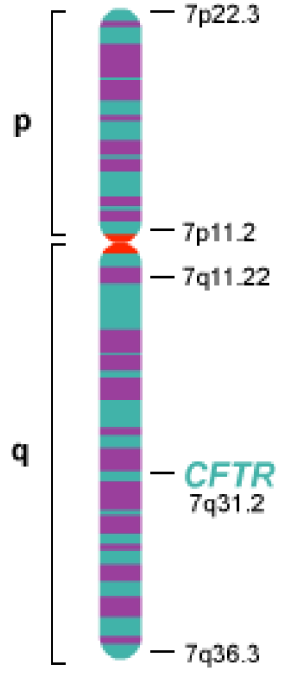
Today CF is the most common fatal genetic disease in Caucasians; attacking the respiratory, digestive, and reproductive systems (CCFF, 2003). While rarely seen in individuals of African or Asian descent, approximately one in 3600 children born in Canada has CF, with one in 25 Canadians carrying the CF gene (CCFF, 2005). Around 3,400 people in Canada attend CF clinics (CCFF, 2005). Even with today's advances in technology and medicine, the median age for life expectancy, as of 2002, is only 37 years (CCFF, 2005).

3.1.2. Pathophysiology.

CF results from the attainment of two copies of a defective autosomal recessive gene, known as *Cystic Fibrosis Transmembrane Conductance Regulator* (CFTR) (Bennett & Peckham, 2002;

CCFF, 2003). Discovered in 1989 by Canadian researchers, CFTR is located on the long arm of Chromosome 7 and is normally responsible for encoding a protein made of 1480 amino acids (Bennett & Peckham, 2002; CCFF, 2003). See Figure 1 below. This protein becomes altered when CFTR is mutated, resulting in the deletion of three nucleotides responsible for producing the amino acid Phenylalanine (Bennett & Peckham, 2002). This amino acid is normally located at position 508 within the protein chain, resulting in the mutation being named delta-F508 (Bennett & Peckham, 2002; see Figure 2 below). While this mutation is present in 66% of all CF cases, there over 1000 mutations which have been detected so far (Sinaasappel et al., 2002).

Figure 1: The Cystic Fibrosis Gene



Name/symbol of Gene Product: cystic fibrosis transmembrane conductance regulator (CFTR)

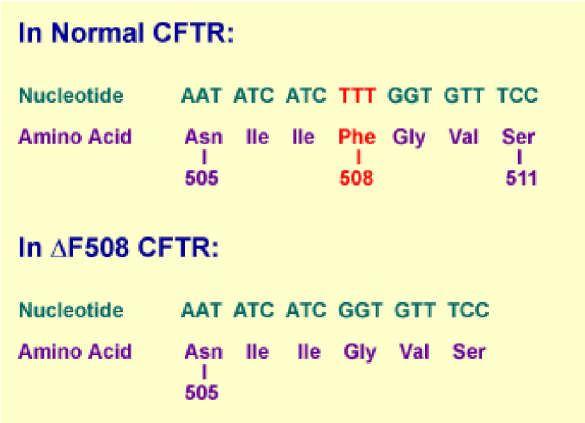
Locus: 7q31.2 The CFTR gene is found in region q31-q32 on the long (q) arm of human chromosome 7.

Size: The CFTR gene's coding regions (exons) are scattered over 250,000 base pairs (250 kb) of genomic DNA. The 27 exons translated into the CFTR protein are interspersed with large segments of noncoding DNA (introns). During transcription, introns are spliced out and exons are pieced together to form a 6100-bp mRNA transcript that is translated into the 1480 amino acid sequence of CFTR protein.

Protein Function: The normal CFTR protein is a chloride channel protein found in membranes of cells that line passageways of the lungs, pancreas, colon, and genitourinary tract. CFTR is also involved in the regulation of other transport pathways. Defective versions of this protein, caused by CFTR gene mutations, cause cystic fibrosis (CF) and congenital bilateral aplasia of the vas deferens (CBAVD).

Source: Gene Gateway- Exploring Genes and Genetic Diseases (n.d.).

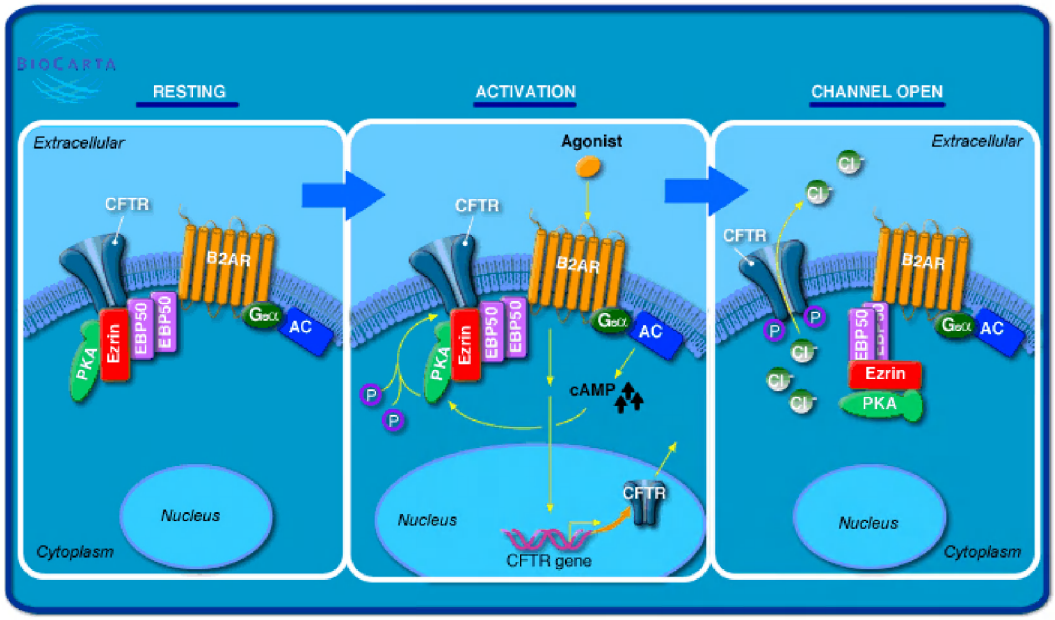
Figure 2: Delta F-508 Mutation Amino Acid Sequence



Source: Gene Gateway- Exploring Genes and Genetic Diseases (n.d.).

Functional CFTR protein forms a chloride channel pore in epithelial cells located in the respiratory, gastrointestinal, hepatobiliary, reproductive, and sweat glands, by binding and hydrolyzing adenosine triphosphate (ATP) (Sinaasappel et al., 2002; Bennett & Peckham, 2002). It also regulates the transport of ions by inhibiting apical sodium transfer across epithelial surfaces, and activating non-CFTR chloride channels (Bennett & Peckham, 2002; see Figure 3). Due to the protein error caused by malfunctioning CFTR, the passage of salts and water across the epithelial cells is not regulated properly, causing the unique salty tasting skin of those with CF, and the production of viscous mucus. This mucus becomes prevalent in the body, particularly in the lungs and digestive system.

Figure 3: Normal Cystic Fibrosis Transmembrane Conductance Regulator Pathway



The defects in cAMP-regulated chloride channel CTFR are believed to be the major cause for cystic fibrosis. Regulation of CFTR protein by the surface receptor beta adrenergic receptor is mediated through the ezrin/radixin/moesin binding phosphoprotein 50 (EBP50), which binds both the C-termini CFTR and b2AR through their PDZ binding motifs. In the resting state, CFTR, b2AR, and EBP50 exist as a macromolecular complex on the apical surface of epithelial cells. Upon agonist activation of the b2AR, the adenulate cyclase is stimulated through the G protein pathway, leading to an increase in cAMP. The elevated concentration of cAMP activates PKA, which is anchored near CFTR via interaction with Ezrin. The phosphorylation of CFTR by PKA disrupts the complex and leads to compartmentalized and specific signaling of the channel.

Source: BioCarta: Charting Pathways of Life. (n.d.).

In the lungs, the altered ion transport creates an imbalance of water, a reduction in the depth of airway surface liquid, and a decrease in ciliary movement (Bennett & Peckham, 2002). The abundance of mucus plays a role in the ability of bacteria, such as *Pseudomonas aeruginosa*, to adhere to the lungs and resist clearance and destruction, as well as making it increasingly harder to breathe (Bennett & Peckham, 2002; CCFF, 2005).

In the digestive system, mucus often blocks the pancreatic duct, reducing the ability of

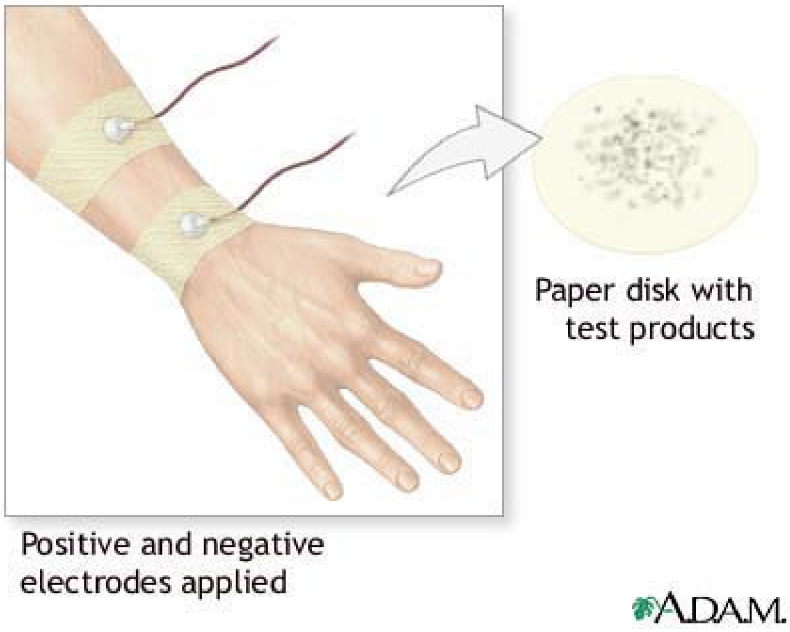
pancreatic enzymes to reach the intestine (Sinaasappel et al., 2002). This pancreatic insufficiency occurs in approximately 85-95% of cases (Sinaasappel et al.; Creveling et al., 1997; Bronstein et al., as cited in Orenstein et al., 2002; Wolfe et al., 2001). When this occurs, pancreatic enzyme replacement therapy (PERT) is initiated, often in pill form, to aid in the digestion and absorption of food (Wolfe et al.). Obstruction of the pancreatic duct can also lead to pancreatic auto-digestion, whereby digestive enzymes get trapped in, and act upon, the pancreas (Sinaasappel et al.).

3.1.3. Diagnosis.

Symptoms of CF include: salty tasting skin, bulky and foul smelling stools, dehydration, a large appetite with poor weight gain, persistent respiratory or intestinal infections, wheezing, and the presence of a persistent cough with thick and often greenish-colored mucus (CCFF, 2003). Parents may notice their infant has a distended abdomen, vomits, and can't move his/her bowels. Infants can also experience failure-to-thrive like symptoms (CCFF, 2003).

The presence of salty tasting skin will help with the diagnosis of CF through the Iontophoretic Sweat Test (see Figure 4), used to measure the amount of salt in sweat (National Library of Medicine, 2006). In this procedure, a small amount of sweat producing liquid is placed on the arm or leg, electrodes are placed over the site, and a small painless current is delivered to induce sweating (National Library of Medicine, 2006). After several minutes, the area is cleaned and sweat is collected for about 30 minutes using gauze or plastic coil tubing.

Figure 4: Iontophoretic Sweat Test



Source: National Library of Medicine, 2006.

From the collected sweat, the amount of sodium and chloride present is determined (National Library of Medicine, 2006).Sweat chloride levels less than 40 millimoles (mmol) per Litre (L) are normal, with levels greater than 60 being indicative of CF (BC HealthGuide Program, 2005). Chloride levels between 40 and 60 mmol/L in infants are reasons to suspect CF (Peckham & Littlewood, 2004).

Genetic screening is another method of diagnosis. If two defective genes are present, the individual has CF (Bennett & Peckham, 2002). This determination can be made in utero, via amniocentesis or chorionic villus sampling (Bennett & Peckham, 2002). One challenge with this method occurs if the individual has a mutation which has not yet been discovered.

3.1.4. Treatment.

Since there is no cure for CF, quality of life is fostered through individualized, on-going, daily treatment. These treatments generally include: efforts to bring up mucus from the lungs and help make breathing easier [manual percussion of the lungs, PEP (Positive Expiratory Pressure) mask therapy, exercise, and/or inhaled medications], taking pancreatic enzymes to aid digestion, taking nutritional supplements and vitamins to improve nutritional status, and taking antibiotics in pill, intravenous (IV), and/or inhaled forms, to ease congestion and protect against, and fight, lung infection (CCFF, 2005).

3.2. Malnutrition and CF

Malnutrition is commonly seen in the CF population, typically presenting as a low body weight and subnormal growth (Sinaasappel et al., 2002). Body weight, and indirectly nutritional status, is dependent upon a balance between an individual's food intake and energy losses. When either of these is out of balance, malnutrition can occur. More specifically, in the CF population, energy imbalances can occur due to a reduced dietary intake, increased energy losses, and increased energy expenditure (Durie & Pencharz, 1989).

3.2.1. Rates of malnutrition.

Studies measuring the dietary intake of CF participants have reported a prevalence of malnutrition between 34% and 59% (Lai et al., 1998; Ionescu et al., 1998, 2000; Dray et al., 2004; Steinkamp & Wiedemann, 2002). Rates can even be high in children as young as two to six years old, as was seen in a study where 19% of this age group were malnourished (Steinkamp & Wiedemann, 2002).Measurements of malnutrition vary between studies and

include Body Mass Index (BMI), Body Mass Index percentile (BMI_p), suboptimal energy intakes, Height-for-Age (HFA), and Weight-for-Age (WFA) percentiles (Lai et al.; Ionescu et al., 2000; Ionescu et al., 1998; Dray et al.; Steinkamp & Wiedemann, 2002).

A study conducted in the United States, evaluated 13,116 children between the ages of birth and 18 years (Lai et al., 1998). Measurements were compared with the National Centre for Health Statistics (NCHS) and Centre for Disease Control's (CDC) growth references (Lai et al., 1998). Results indicated that the mean percentile for HFA was 30, and for WFA was 20 (Lai et al.). Malnutrition was seen in 34% of the adolescent participants, with stunting occurring in 19% of the males and 29% of the females, aged 11 to 14, and 34% and 28% of those aged 15 to 18, respectively (Lai et al.). Here, stunting was defined according to a HFA of less than 5% and/or a HFA of less than 90% of a reference median (Lai et al.). The Cystic Fibrosis Foundation's (US) 2001 patient registry indicated that 18% of children fell below the CDC's 5th percentile for weight and 16% fell below the 5th percentile for height.

3.2.2. Reduced dietary intake.

A poor dietary intake is well documented in the CF population, especially in males (Collins, O'Loughlin, & Henry, 1998). There are many reasons for a poor dietary intake including:

- Anorexia: which may be caused by respiratory involvement, depression, and esophagitis;
- Feeding disorder (neonatal tube feeding);
- Depression;
- Interaction with parents;
- Esophagitis: which may be due to reflux that causes pain, anorexia, and vomiting;
- Distal Intestinal Obstruction Syndrome (DIOS; meconium ileus equivalent): causes

recurrent crampy abdominal pain, which is made worse by eating;

- Iatrogenic fat restriction;
- Decreased appetite from drugs or infection;
- Inflammation;
- Lack of general well being;
- Extrahepatic biliary obstruction;
- Cholangitis;
- Advanced liver disease; and/or
- Severe constipation (Sinaasappel et al., 2002; Pencharz & Durie, 1993).

When measuring the dietary intake of 40 adults with CF, it was found that 21 had a low caloric intake when compared to recommendations (Ionescu et al., 2002). The mean energy intake for the whole group was 2354.9 calories (Cal) a day, of which 38.8% was obtained from fat (Ionescu et al.). Caloric intake values vary between studies, from as low as 686 to as high as 5010 Cal a day, with an average intake of around 2200 (Steinkamp, Demmelmair, Ruhl-Bagheri, von der Hardt, & Koletzko, 2000; Marin et al., 2004; Ionescu et al.; Walkowiak & Przyslawski, 2003; Stark et al., 1997; Collins et al., 1998). One study found a difference in intake between genders, with males consuming an average of 131% and females consuming 107% of the World Health Organization's (WHO) daily recommendations (Collins et al.). While more than half of the participants of one study consumed caloric intakes in the recommended amounts, CF participants were significantly shorter and lighter when compared to control participants (Stark et al.).

3.2.3. Increased energy loss.

Energy deficits can occur when the body's energy needs outweigh the individual's energy intake (Pencharz & Durie, 1993). Energy is typically lost in the CF individual through malabsorption and maldigestion. Those with CF who are pancreatic insufficient, compensate for a lack of digestive enzymes by taking enzyme replacements. These replacements vary with respect to their enzyme content and dissolution in relation to pH and duodenal fluid (Atkinson, as cited in Sinaasappel et al., 2002). However, even at the recommended dose [10,000 International Units (IU) lipase per kilogram (kg) of body weight]; symptoms of malabsorption may still be seen (Durie, Kalnins, & Ellis, 1998).

One of these symptoms is a loss of energy in the stool. The amount of energy typically lost by healthy adults in their stool is around 5% of their total energy intake (Murphy et al., 1991). However, Murphy et al. (1991) determined that stool energy losses in 20 participants with CF, aged five to 25 years, accounted for around 10.6% of the individual's daily calorie intake. Around 41% of the energy lost in the stool was due to malabsorption of lipids, with another 30% being caused by bacteria (Murphy et al.).

Other problems may also affect the absorption of nutrients. It has been speculated that the pancreas of those with CF is unable to produce adequate bicarbonate, due to the altered water and electrolyte transport (Gaskin, Durie, Corey, Wei, & Forstner, 1982). This was indeed seen by Gaskin et al. (1982), in a study involving 62 CF participants. Such a defect in bicarbonate production, possibly caused by the genetic mutation, leads to a decrease in the intestinal pH, impacting absorption (Gaskin et al., 1982). Most new enzymes replacement tablets are coated with an acid-resistant material and may not dissolve in the proximal area of the intestine when

there is an imbalance of acid (Sinaasappel et al., 2002). As a result, fatty acids remain in the oil phase and are not properly absorbed (Sinaasappel et al.). A low intestinal pH can also denature pancreatic lipase (Pencharz & Durie, 1993), the enzyme responsible for digesting fats (Sinaasappel et al.).

3.2.4. Increased energy expenditure.

A person's Total Energy Expenditure (TEE) is comprised of three parts: 60-70% is lost due to the Resting Energy Expenditure (REE), 10-25% due to physical activity, and 10% due to diet induced thermogenesis (Sinaasappel et al., 2002). The presence of CF places higher energy demands on the individual.

The REE of those with CF has been found to be anywhere from four to 80% (typically 20-50%) higher than that of healthy peers (Pencharz, Hill, Archibald, Levi, & Newth, as cited in Pencharz & Durie, 1993; Sinaasappel et al., 2002; UK Cystic Fibrosis Trust Nutrition Working Group, 2002; Creveling et al., 1997; Marin et al., 2004). Indirect calorimetry, performed on malnourished participants with moderate to advanced lung disease, was used to asses the relationship between heart rate and energy expenditure (Pencharz et al., cited in Pencharz & Durie, 1993). Results indicated that participants with CF were found to have an REE 25-80% higher than those of healthy individuals, of the same age, sex, and size (Pencharz et al., cited in Pencharz & Durie, 1993). Another study found that the REE, in participants without a respiratory infection, was within 95 – 153% of the normal range (Vaisman, Pencharz, Corey, Canny, & Hahn, as cited in Pencharz & Durie, 1993).

This increase in REE is thought to be caused by the status of the lungs, and perhaps by the genetic defect itself (Pencharz & Durie, 1993). Energy demands increase as the lungs become

progressively destroyed. Air trapped in the lungs, which may account for up to 10% of the REE, causes them to become hyperinflated and makes it difficult for the diaphragm to work properly (Ionescu et al., 1998; Elborn & Bell, 1996). The work of breathing will continue to increase as the lungs worsen, demanding more and more energy from the individual (Pencharz et al., as cited in Pencharz & Durie, 1993). During exacerbations of infection, the nutritional status is negatively affected. While this can be helped by antibiotics, end stage disease often causes a cycle of exacerbation, pulmonary and systemic inflammation, and a decline in body weight (Elborn & Bell, 1996).

REE can also be increased by medications used to treat CF. The beta-agonist, salbutamol (*Ventolin*®), can increase REE by ten percent for up to three hours after use (Vaisman et al., as cited in Pencharz & Durie, 1993).

3.2.5. Additional factors.

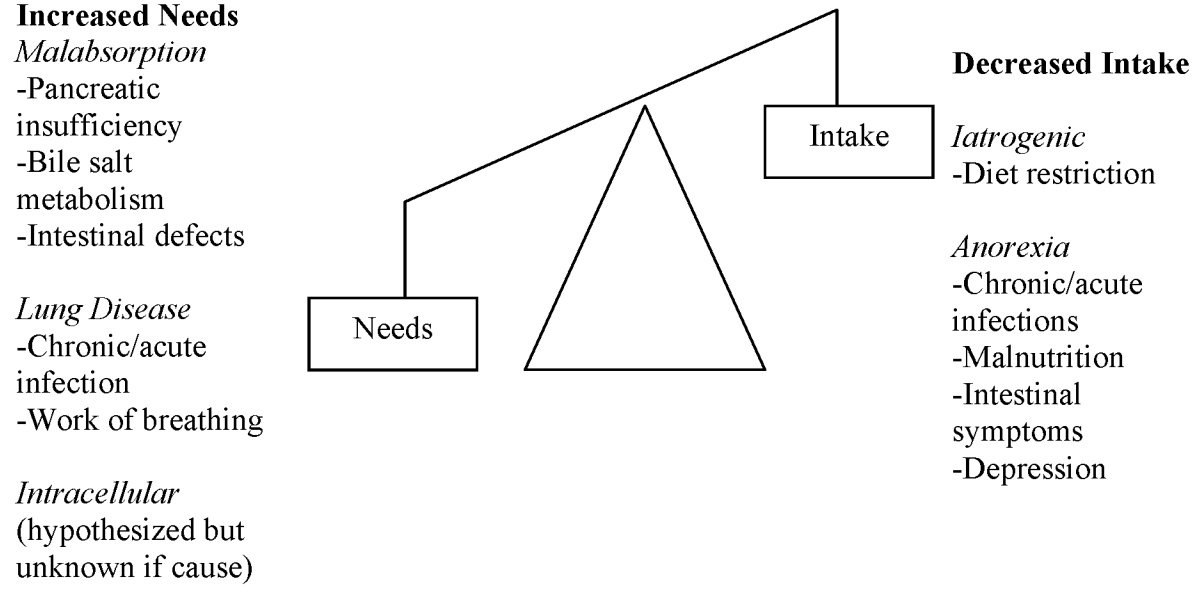
Secondary complications may arise in the CF population, placing even greater energy demands on the body. Glucose intolerance develops in around 50-70% of CF cases and around ten to 30% will develop cystic fibrosis related diabetes mellitus (Creveling et al., 1997; Littlewood, 2002). If untreated, energy may be lost as glucose in the urine (Pencharz & Durie, 1993).

CF results in multi-organ involvement. Between two and 37% of those with CF have some form of liver involvement, with around 10% developing cirrhosis (Sinaasppel et al., 2002). With signs of cirrhosis, the presence of bile salts, required for the digestions of fats, may be impaired. Such deficiencies will compromise lipolysis, leading to a decrease in fat absorption of up to 50% (Austgen, Bowen, & Rouge, 2001; Pencharz & Durie, 1993; Sinaasppel et al.).

During periods of inflammation, which are common in CF, the cytokine Tumor Necrosis Factor-α (TNF) will be involved (Elborn, Cordon, Western, MacDonald, & Shale, 1993). Elevated levels of TNF are seen in those with CF and have been associated with body wasting, and an increase in REE (Elborn et al., 1993). These high levels may have negative consequences such as malnutrition, leading to an increase in morbidity and mortality (Elborn et al.).

While it is known that CFTR is expressed in the lungs and pancreas, a study on rat brains found it to be localized and expressed in the areas of the hypothalamus, thalamus, amygdala, and limbic system (Mulberg et al., 1995). These areas of the brain play a large role in food intake, energy expenditure, and metabolic rate (Mulberg et al.). If CFTR is similarly expressed in the brains of humans, this may lead to common CF symptoms such as an increased REE, growth failure, and malnutrition. Figure 5 depicts the causes of energy imbalances in CF.

Figure 5: Causes of Energy Imbalances in Cystic Fibrosis



Source: Durie & Pencharz, 1989.

3.3. Nutrient Deficiencies

3.3.1. Essential fatty acids.

Essential Fatty Acid (EFA) levels in the CF population are often sub-optimal, even in those who are well nourished, consume a high fat diet, and show no signs of malabsorption (Strandvik, as cited in Sinaasappel et al., 2002; Steinkamp et al., 2000; Roulet, Frascarolo, Rappaz, & Pilet, 1997). A deficiency of EFA, usually defined according to a decrease in linoleic and possibly alpha-linolenic acid, has been seen in the plasma and tissues of those with CF (Strandvik, as cited in Sinaasappel et al.). There have also been reports of decreased levels of omega-3 long-chain Polyunsaturated Fatty Acids (PUFA), such as eicosapentaenoic and docosahexanoic acid (Strandvik, Gronowitz, Enlund, Martinsson, & Wahlstrom, 2001). An altered EFA profile is associated with altered membrane and cellular functions, skin lesions, and an altered immune, renal, hepatic, and respiratory function (Steinkamp et al., 2000).

There are a few proposed reasons for EFA deficiencies in the CF population. It has been shown that study participants who have one copy of the common delta F508 genetic mutation (instead of two) have significantly lower serum concentrations of linoleic acid when compared to other CF and control participants (Strandvik et al., 2001). The decreased level of EFA may also be linked to the basic defect in CF, micronutrient deficiencies, a defect in the inclusion of fatty acids in the cell membrane, a disorder of lipoprotein metabolism (Roulet et al., 1997), a defective regulation resulting in an increased turnover of arachidonic acid from phospholipids (Carlstedt-Duke, Bronnegard, & Strandvik, 1986), enhanced oxidative stress and peroxidative destruction of PUFA during chronic infection, a low body weight, and/or a negative energy balance (Sinaasappel et al., 2001, 2002).

3.3.2. Vitamins.

Vitamins play an important role in the body and it is essential that levels are maintained in individuals with CF. Of particular importance are the fat soluble vitamins A, D, E, and K. Deficiencies of the first three vitamins have presented as early as up to two months of age in CF patients (Feranchak et al., 1999). The majority of deficiencies are remedied with the initiation of pancreatic supplements, if needed, and with vitamin supplementation (Feranchak et al.).

Vitamins A and E are antioxidants which play an important role in combating the high oxidative damage created by chronically inflamed lungs (Sinaasappel et al., 2002). As a result, it is recommended that all antioxidant levels, including those of water soluble vitamins, be maintained (Sinaasappel et al.). Low serum levels of vitamin A are common in the CF population, as well as low plasma and tissue carotenoid levels (Sinaasappel et al.; Lancellotti, D'Orazio, Mastella, Mazzi, & Lippi, 1996). These decreased levels may be more pronounced in those who are pancreatic insufficient, which unfortunately are not usually remedied by taking enzymes and/or supplements (Sinaasappel et al.; Lancellotti et al., 1996). This suggests that there may be a disturbance in the use of hepatic vitamin A stores, the amount of supplementation may be inadequate, or there may be a problem handling retinol in the gastrointestinal tract (Sinaasappel et al.; UK Cystic Fibrosis Trust Nutrition Working Group, 2002). It is recommended that those who are pancreatic insufficient be given supplements of vitamin A at doses of between 4000 and 10,000 IU, dependent upon age (Peters & Rolles, 1993; Sinaasappel et al.).

Similar to vitamin A, while the majority of new diagnoses show low levels of vitamin E, levels may not be replenished by using enzyme replacement and vitamin supplementation

(Lancellotti et al., 1996; Feranchak et al., 1999). Due to this problem, it is important to monitor vitamin E status periodically (Feranchak et al.). For those with chronic cholestatic liver disease, a potential cause of lower vitamin E status is the way their bodies handle alpha tocopherol, suggestive that a decrease in bile reaching the duodenum may play a role in the deficiency of the vitamin (Winklhofer-Roob, Shmerling, Schimek, & Tuchschmid, 1992; Ortiz Zein & Lindor, 2000).

It is still unknown whether supplementation of vitamin E results in a decreased amount of oxidative damage to the lungs (Sinaasappel et al., 2002), but there has been evidence of a positive correlation between vitamin E status and lung function, as assessed by Forced Expiratory Volume in one second (FEV_1) (De Vriese, Robberecht, De Gussem, & Christophe, 1999). Therefore, while clinical deficiency symptoms of vitamin E are rare in CF patients (Sinaasappel et al.), ensuring that vitamin E levels are normal will help prevent premature lipid peroxidation (Winklhoefer-Roob et al, 1992). Levels of supplementation in the literature vary in amount and form, with studies indicating the use of 100 milligrams (mg) a day to 250, and the use of the ursodeoxycholic acid form of vitamin E (Peters & Kelly, 1996; Feranchak et al., 1999; Thomas, Bellamie, & Geddes, 1995).

While not an antioxidant, 22.5% of newborns diagnosed with CF have been found to have low levels of vitamin D during the first two months of life (Feranchak et al., 1999). It has been hypothesized that vitamin D deficits may be caused by low serum levels of vitamin D binding protein, suggestive of a transport problem (Coppenhaver et al., 1981). While suboptimal serum levels of vitamin D are common in CF patients, signs of vitamin D deficiency, such as rickets, are rare (Sinaasappel et al., 2002; UK Cystic Fibrosis Trust Nutrition Working Group, 2002).

This suggests that signs of nutrient deficiencies may be harder to detect in patients with CF, and that reliance upon biochemical assessment is important. Doses between 400 and 800 IU usually result in normal serum levels at the lower end of the optimal range; however, some patients may require up to 2000 IU a day (Peters & Rolles, 1993; Sinaasappel et al.). Supplementation is also recommenced for those who live in areas of the world with inadequate sunlight exposure (Sinaasappel et al.).

Unlike vitamin D, symptoms of vitamin K deficiency are more easily seen. Vitamin K is a co-factor in the carboxylation of prothrombin (used in blood clotting), and in bone formation (Whitney & Rolfes, 1999, p. 354; UK Cystic Fibrosis Trust Nutrition Working Group, 2002). People with CF are at a risk of being deficient in vitamin K due to fat malabsorption, bile salt deficiency, liver disease, and antibiotic use (Durie & Pencharz, 1989). A new method of assessing vitamin K status called PIVKA II (proteins induced by vitamin K absence) measures the amount of decarboxylated prothrombin and high levels may be indicative of a vitamin K deficiency (Rashid et al., 1999).

Low vitamin K, as assessed by PIVKA II presence in the blood, is common in CF patients who are pancreatic insufficient (78%), have liver disease (100%), and even those who are pancreatic sufficient (33%) (Rashid et al., 1999). For this reason, vitamin K supplementation should be given to those with CF who also have liver disease, and/or show an increased clotting/prothrombin time (UK Cystic Fibrosis Trust Nutrition Working Group, 2002). Studies have found that receiving a single dose of five mg of vitamin K a week did not prevent subclinical deficiencies, indicating that perhaps five mg is not a high enough supplementation dose (Beker et al., 1997). However, the best amount of vitamin K supplementation is still controversial and undetermined (Beker et al.).

Water soluble vitamins usually do not pose a problem in those with CF (Sinaasappel et al., 2002; UK Cystic Fibrosis Trust Nutrition Working Group, 2002). Vitamin B_{12} may be an exception in those with pancreatic insufficiency, which is lessened with enzyme replacement therapy (Sinaasappel et al.). It is recommended that those who have had extensive surgery for meconium ileus, need lifelong Total Parenteral Nutrition (TPN), and that individuals who have had a resection of the terminal ileum should take supplements of B_{12} (Sinaasappel et al.).

3.3.3. Minerals.

There are only a few minerals of particular importance to the CF population. Sodium and chloride are the most important ions in extracellular fluid (Legris, et al., 1998). The average adult loses 500 mg of sodium in sweat per day. This can increase ten times in people with CF, especially during exercise, or in hot weather (Sinaasappel et al., 2002). These high losses can lead to hyponatraemia and alkalosis, and it is recommended that extra salt be taken during periods of excessive sweating (Sinaasappel et al.).

Another mineral of concern is iron, since deficiencies are common in CF due to an inadequate dietary intake, malabsorption, chronic infection, and blood loss (Sinaasappel et al., 2002). Pancreatic enzymes may be responsible for impaired iron absorption and should not be taken with supplements (Zempsky, Rosenstein, Carroll, & Oski, as cited in Sinaasappel et al.).

Lastly, the micronutrient zinc may be lowered in CF patients (Easley et al., 1998; Sinaasappel et al., 2002). Zinc absorption is thought to be affected by fat malabsorption, causing zinc to form complexes with fat and phosphorus (Krebs, Sontag, Accurso, & Hambidge, 1998). Enzyme therapy can improve zinc absorption (Easley et al.).

3.4. Effects of Nutrition on Physiology

3.4.1. Growth.

Infants born with CF often have a lower birth weight than healthy infants (Mearns, as cited in Sinaasappel et al., 2002). The average CF birth weight is 3.18 kg for males and 3.04 kg for females, as compared to an average healthy birth weight of 3.37 kg and 3.25 kg, respectively (Mearns, as cited in Sinaasappel et al.). Growth during the infancy period can be normal if chest infections are prevented or treated, and malabsorption problems are dealt with (Simmonds, Wall, Wolfe, & Littlewood, as cited in Sinaasappel et al.). This growth can be achieved via breast milk or formula (Holliday et al., 1991). Canadians who have CF have almost normal weight and height increases throughout childhood. If there is a lag in growth during infancy, it is possible to catch-up during childhood (Corey, McLaughlin, Williams, & Levinson, 1987).

3.4.2. Lung function.

The nutritional status of those with CF impacts lung function. Having a normal weight has been shown to result in a smaller decline in lung function when compared to those who are malnourished, often independent of infection (Steinkamp & Wiedemann, 2002; Konstan et al., 2003). Steinkamp & Wiedemann (2002) identified individuals as malnourished if their weight-for-height (WFH) was less than 90% of predicted. Malnourished adolescents, who had a decrease in WFH of five percent or more over one year, experienced a loss in FEV_1 of 16.5%; whereas those who gained weight showed an increase of 2.1% (Steinkamp & Wiedemann, 2002).

The impact of malnutrition may be long term; two groups of children with CF were evaluated at age three and six, one group having a weight-for-age (WFA) below the 5^{th} percentile and the other with a WFA above the 75^{th}. Those in the low WFA group scored lower on lung function

tests that those in the high WFA group, three years later (Konstan et al., 2003). This decline in function was more noticeable in children with a low body WFA as opposed to height-for-age (HFA) (Konstan et al.). On a more positive note, the lungs may benefit from an increased nutritional status, leading to an increase in FEV_1 when weight increases from below to above the tenth percentile (Konstan et al.).

Another consequence of generalized weight loss is a loss of fat free mass (FFM), including lung musculature (Ionescu et al., 1998). A loss of FFM may also be caused by physical inactivity, malabsorption, steroid therapy, reduced sex hormone levels, increased REE, and hypoinsulinemia when diabetes mellitus is present (Ionescu et al., 2000). It has been seen that as the body composition of CF patients decreases, inspiratory muscle function can decrease as well (Ionescu et al., 1998). A study of 40 adults with CF who were colonized with the bacterium *Pseudomonas aeruginosa,* found that 24 participants with a low FFM had a lower mean FEV_1 (53.9%) than those with a normal FFM (FFM > fifth percentile) (72.4%) (Ionescu et al., 2002). A loss of FFM has also been related to the number of pulmonary exacerbations in the previous year, with a mean of 4.7 days versus 2.7 days for normal FFM patients (Ionescu et al., 2002). As well, the number of exacerbations was related to lung function, as predicted by FEV_1 values in the previous year (Ionescu et al., 2002).

As can be seen from this data, body composition, particularly muscle mass, plays a significant role in lung function. Bacterial infections, which are common in CF, can create a cycle of malnutrition and lung deterioration (Ionescu et al., 2002; Pencharz & Durie, 1993). When FFM is lost, the muscles of the lungs are impaired, leading to increased pulmonary problems, more inflammation, and further deterioration of lung function (Ionescu et al., 1998;

Pencharz & Durie, 1993). A decrease in lung function increases the energy needed to breathe and, if the nutritional status of the person is compromised, further protein catabolism results from the negative energy balance (Ionescu et al., 2002; Pencharz & Durie, 1993). It has been shown that when infections are treated with antibiotics, the inflammatory response and rate of catabolism declines (Ionescu et al., 2000). However, chronic infection is common in CF.

3.4.3. Bone health.

Osteoporosis/osteopenia is a condition characterized by a decrease in bone mineral density, making bones porous and fragile (Whitney & Rolfes, 1999, p. 15 [Glossary]). The World Health Organization identifies osteopenia according to a bone mineral density (BMD) value between one and two and a half standard deviations (SD) below the mean for young, healthy adults (WHO Scientific Group, 2003). Osteoporosis is identified according to a BMD value of two and a half standard deviations, or more, below the mean (Elkin et al., 2001).

Rates of osteopenia or osteoporosis in the CF population have been documented as between 42 and 60% (Conway et al., 2000; Fok, Brown, Zuberbuhler, Tabak, & Tom, 2002). This secondary health problem develops in the CF population due to factors such as: malnutrition and malabsorption, an inadequate nutrient intake, increased energy requirements, the degree of disease severity, reduced physical exercise, an increase in bone metabolism, reduced sex hormone levels, hypoinsulinemia in cases of diabetes mellitus, and low body weight (Bachrach, Loutit, Moss, & Marcus, 1994; Ionescu et al., 2000, 2002). The presence of a low body weight is one of the most consistent predictors of a low BMD (Bachrach et al., 1994; Turner et al., 2001). It has been shown that a low fat-free mass, as opposed to fat mass, in females who have disordered eating behaviors, is the strongest independent factor negatively affecting BMD (Bachrach et al., 1994; Turner et al., 2001).

The presence of low bone density has been found in the CF population as early as age five to eight years (Fok et al., 2002). From then on, it seems that BMD rates can continue to decline at a rate of approximately one SD for every six to eight years of age (Fok et al.). The status of the lungs and the presence of chronic inflammation can lead to an inflammatory and catabolic response in the body, even during periods of clinical stability (Elborn et al., 1993). Catecholamines, proinflammatory cytokines, and stress hormones such as cortisol, can lead to a breakdown of protein in bone (Ionescu et al., 2000; Elborn et al.).

As described earlier, nutritional status has an effect on the lungs and thus in this manner, indirectly effects bones. One study documented that participants with a BMI below the fifth percentile had a mean BMD of -2.5 (Fok et al., 2002). Research has found that the CF population has a higher rate of bone metabolism than healthy individuals stressing the importance of body weight maintenance (Ionescu et al., 2002). A loss of FFM, in addition to negatively impacting the lungs, can have a negative impact on bone mineral density (Ionescu et al., 2002). Pseudouridine, an indicator of ribonucleicacid (RNA) catabolism, was found to be present in the urine of CF individuals, indicating increased bone turnover (Ionescu, Nixon, Buss, Routledge, & Shale, 1999). The excretion of this bone metabolite was shown to be related to lung disease status, whereby those with a low FFM had a significantly lower mean FEV_1 percentage (53.9%) when compared to those with a normal FFM (72.4%) (Ionescu et al., 2002).

To try to keep the bones healthy, it is recommended that those with a BMI in the 25^{th} percentile, or below, begin some form of aggressive nutritional therapy, to reduce the risk of developing osteoporosis (Fok et al., 2002). As well, it is recommended that those with CF consume adequate amounts of vitamin D and calcium from the diet or other sources. A

deficiency of vitamin D, combined with malabsorption, will affect the absorption of calcium (Conway, 2001). Also, since vitamin D itself is important for bone health, it is recommended that patients take a vitamin D supplement if they do not get adequate sunlight.

3.4.4. Puberty.

During adolescence, in general, energy requirements increase due to growth, puberty, and a possible increase in physical activity (UK Cystic Fibrosis Trust Nutrition Working Group, 2002; Pencharz & Durie, 1993). This is also true for those with CF and, while they may not have physical activity levels similar to those of their healthy peers, malnutrition at this time can lead to a delayed puberty (UK Cystic Fibrosis Trust Nutrition Working Group; Pencharz & Durie, 1993).

It is hypothesized that CFTR may play an indirect role in pubertal delay due to the fact that CFTR messenger ribonucleic acid (mRNA) has been found to be expressed in the uterus, cervix, and fallopian tubes (Tizzano, Silver, Chitayat, Benichou, & Buchwald, 1994; Johannesson et al., 1997).While CFTR is not expressed in the ovaries, they contain cells with insulin receptors, which play a role in the production of gonadal steroids (Johannesson et al.). The gonadotrophic activity of insulin occurs through: direct effects on the steroidogenic enzymes, alteration of follicle stimulating or luteinizing hormone receptor numbers, or synergistic action with both of these hormones (Poretsky & Kalin, as cited in Johannesson et al.). Glucose intolerance develops in around 50-70% of CF cases and around ten to 30% will develop cystic fibrosis related diabetes mellitus (Creveling et al., 1997; Littlewood, 2002). When an Oral Glucose Tolerance Test was given to 17 females, those with disordered values were significantly older than those with normal values (15.8 ± 1.7 years versus 14.3 ± 0.9 years, respectively) (Johannesson et al.).

Genotype may also play a role in the delay of puberty of CF adolescents (Johannesson et al., 1997). Those who have two copies of the typical mutated gene (homozygotes), delta F508, show a significant delay when compared to those who do not contain two copies of this mutation (15.2 ± 1.9 years versus 14.7 ± 0.9 years, respectively) (Johannesson et al.). In another study, it was found that the REE of homozygotes was around 10- 15% higher than that of individuals with other mutations (O'Rawe et al., 1992). This increased energy requirement could be a possible explanation for the cause of delayed menarche (Johannesson et al.). Also, as stated previously, CFTR mRNA has been found to be expressed in areas of the rat brain such as the hypothalamus, thalamus, and amygdala, which are important sites for the regulation of basal metabolism, glucose control, and sexual maturation (Mulberg et al., 1995). If expression is similar in humans, this may be another cause of delayed puberty.

A group of 30 participants with CF, male and female, were studied to determine the rate of pubertal development (Aswani, Taylor, McGaw, Pickering, & Rigby, 2003). It was found that the age at which a growth spurt occurred, or the attainment of peak height velocity, was significantly delayed in both sexes versus normal controls (Aswani et al., 2003). However, final heights were not significantly different between the CF and control populations, with 52% of the CF group reaching at least the same final height percentile when compared to their parent's height (Aswani et al.). Therefore, while children with CF may enter puberty later, they may continue to grow longer than their peers.

In females with CF, menses may be delayed by as much as 4.2 years when compared to healthy age mates by 4.4 years when compared to their mothers (Johannesson et al., 1997). The Frisch-Revelle hypothesis states that a weight between 46 and 48 kg must be met before

menarche can occur (Frisch, 1969; Frisch & Revelle, 1971). A minimal body fatness level of 17% has also been found to be required for initiation of menstrual cycles, and a level of 22% will maintain regular cycles (Frisch, 1976). However, it has been found that some females with CF will begin menstruating at a more typical age of 13, despite the fact that the critical weight is not met (Johannesson et al.).

A delay in puberty is not only a female phenomenon. CFTR has been found to be expressed in the epididymis and vas deferens in males with CF (Tizzano et al, 1994). As a consequence of the disease, most males with CF show atrophy or obstruction of the epididymus, vas deferens, and seminal vesicles; others are born without vas deferens and may have partially developed seminal vesicles (Tizzano et al.).

A group of 33 CF males, aged 13 to 17 years, participated in a study which looked at the relationship between pubertal delay, nutritional status, and lung function (Boas et al., 1998). Tanner staging, a method of determining the stage of pubertal development based on physical signs, was used to assess the male participants (Tanner, 1962). As well, serum and salivary testosterone levels were assessed (Boas et al.).

According to the Tanner staging, delayed puberty is assessed by a failure to reach the expected stage by the age at which 95% of healthy children have reached it (Tanner, 1962). Out of the sample, 20 showed a delay in puberty (Boas et al., 1998). The average height, weight, BMI, mid-arm muscle area, and FEV_1 differed between those who were delayed and the 13 who were not (Boas et al.). It was determined that males were more likely to have a delay in pubertal development if they had: a weight $\leq$ tenth percentile, a height $\leq$ the 25^{th} percentile, and a BMI $\leq$ 25^{th} percentile (Boas et al.). FEV_1 scores below the 60^{th} percentile were associated with a delay

in puberty when compared to those with a score above the 60th percentile (Boas et al.).

On the average, male genital development is delayed by 1.6 years, compared to that of healthy peers; and females are delayed by two years (Sawyer, Rosier, Phelan, & Bowes, 1995). Damaged self-esteem and altered body image perception can result from this delay (Sawyer et al., 1995).

3.5. Nutritional Management

3.5.1. History.

As early as the 1940's, nutrition was seen as an important aspect of the care and treatment of CF (Littlewood, 2002). Diets were recommended to be high in protein and carbohydrates, low in fat, and supplemented with vitamin A, due to the belief that a deficiency of vitamin A was partly to blame for the condition (Littlewood, 2002). Pancreatic extracts, available since the 1930's, were also given with food. Dorothy Anderson noted that such dietary therapy should be initiated as early as possible, to allow for the best outcome (Littlewood, 2002). Patients who were fed six to eight grams (g) of protein/kg/day, in caseine hydrolysate forms, showed good growth rates (Littlewood, 2002). The only fat recommended was from one to two eggs a day and fish oil supplements (Littlewood, 2002). The reasoning behind the low fat diet was that an intake reduced in fat would be easier on the bowels and reduce unwanted symptoms of malabsorption and maldigestion (Corey et al., 1987).

In the 1960's, it was recommended that infants be fed a low fat, high protein formula, and be started on solids at two months (Littlewood, 2002). A tablespoon of casein hydrolysate and simple sugars was often added to formulas (Littlewood, 2002). Water soluble vitamins were

given in twice the amount recommended, along with 50 mg of vitamin E for infants and 100 mg for children, and five mg of vitamin K two times a week if infants were on antibiotics (Littlewood, 2002). Older patients were fed a high protein, low fat diet. Newer pancreatic enzymes were available now, such as Cotazyme (Littlewood, 2002).

The 1970's was a decade of major change for nutrition therapy in CF. A more liberal diet was adopted, with older children eating 30-40 g of fat a day (Littlewood, 2002). It was also recommended that 200 Cal/kg/day, and four to five g protein/kg/day, be consumed. *The Allan Diet,* created by Dr. Allan a pediatrician from England, used a supplement of beef serum protein hydrolysate, glucose polymers, and medium chain triglycerides to treat those with CF. Despite the fact that medium chain triglycerides have been shown to reduce the symptoms of malabsorption, such a restrictive diet was short lived, and clinicians concluded that it should be used in specific cases only (Littlewood, 2002).

Also in the 1970's, a Toronto doctor (Crozier) felt it was not right to restrict the diet of people with CF to the extent that was common (Corey et al., 1987; Crozier, 1974). As early as 1972, Crozier began to recommend that patients eat a diet high in saturated fats such as whole milk, eggs, butter, and animal products (Corey et al.; Crozier, 1974). This increase in fat was matched by an increase in pancreatic enzymes.

Research determined that these new dietary recommendations could possibly increase survival rates. A study comparing the Toronto clinic with one in Boston, showed a correlation between longevity and nutritional status (Corey et al., 1987). The median age of survival of patients at the Boston clinic was 21 years, verses 30 years in Toronto (Corey et al.). While pulmonary function results did not differ significantly between the two clinics, individuals in the

Toronto clinic were taller than those in Boston. Also, males attending the Toronto clinic were heavier than males attending the Boston clinic (Corey et al.). The difference in the two clinics was found to exist in their dietary recommendations. It was then determined that Dr. Crozier's recommendations led to an improved body weight and better health status in those with CF (Corey et al.).

3.5.2. General recommendations.

The goal of nutritional management for people with CF, be they infants or adults, is to help facilitate normal growth, development, and maintenance, leading to an increased quality of life. Currently, it is generally recommended that the CF population consume 120 to 150% of calories recommended for similar healthy peers (Roy, Darling, & Weber, as cited in Pencharz & Durie, 1993; Pencharz et al., as cited in UK Cystic Fibrosis Trust Nutrition Working Group, 2002; Sinaasappel et al., 2003; Creveling et al., 1997). Carbohydrate and protein recommendations are similar to those for the general population; however, fat is a major source of energy and should be increased to 30-40% of daily calories, due to malabsorption (Pencharz et al., as cited in Pencharz & Durie, 1993; Marin et al., 2004). This dietary strategy, rooted in the work of Dr. Crozier, results in an overall increase in the nutritional status of CF patients (Pencharz et al., as cited in Pencharz & Durie, 1993).

3.5.3. CF adolescent nutrition.

During adolescence, a time when protein and energy requirements are at their peek for those with or without CF, nutrition therapy for those with CF should maintain or restore a normal nutritional status and growth patterns (UK Cystic Fibrosis Trust Nutrition Working Group, 2002). As described earlier, a positive nutritional status can help improve the lungs, prevent

osteoporosis/osteopenia, and prevent pubertal delay (UK Cystic Fibrosis Trust Nutrition Working Group, 2002).

3.5.4. Invasive nutritional support.

When those with CF are unable to meet their energy needs orally, the use of supplements may be helpful (UK Cystic Fibrosis Trust Nutrition Working Group, 2002). It is recommended that oral supplements be prescribed to increase energy intake in children and teens whose WFH is between 85 and 89%, who have experienced a weight loss over four to six months, or who have leveled off in weight gain over six months (Sinaasappel et al., 2002). Supplements should not be used as a meal replacement, but in addition to regular food (UK Cystic Fibrosis Trust Nutrition Working Group, 2002).

If nutritional status does not improve with oral supplement use, invasive methods of energy supplementation, such as enteral feeds, may be recommended. In general, enteral nutrition is recommended in children after the use of oral supplements and when the child has a WFH below the 85th percentile, a weight falling two percentiles, or when there has been no weight gain for a period of six months (UK Cystic Fibrosis Trust Nutrition Working Group, 2002; Sinaasappel et al., 2002).

Supplemental feedings lead to positive changes in body fat and muscle, linear growth, and the ability to take part in every-day activities (Levy, Durie, & Pencharz, as cited in UK Cystic Fibrosis Trust Nutrition Working Group, 2002). Nasogastric feedings can be used for short durations during exacerbations, as a boost for growth, or as a trial before gastrostomy (UK Cystic Fibrosis Trust Nutrition Working Group, 2002). Users are often trained to insert and remove tubes and usually feed at night (UK Cystic Fibrosis Trust Nutrition Working Group,

2002). Some complications that can develop include nasal discomfort, tube blockages, aspiration, and nausea/bloating/vomiting (UK Cystic Fibrosis Trust Nutrition Working Group, 2002).

Gastrostomy feedings are usually used for long-term supplementation (UK Cystic Fibrosis Trust Nutrition Working Group, 2002). Feedings can be done overnight and throughout the day, as needed. Tubes are usually inserted endoscopically, under sedation or general anesthetic and can result in complications such as tube blockages, pain from insertion, nausea, bloating, vomiting, and less commonly peritonitis (UK Cystic Fibrosis Trust Nutrition Working Group, 2002).

As a last resort, Total Parenteral Nutrition (TPN) is used usually for short durations, and/or post- surgery (UK Cystic Fibrosis Trust Nutrition Working Group, 2002). TPN has little added benefits over enteral nutrition, is often more expensive, and has a greater risk of complications (UK Cystic Fibrosis Trust Nutrition Working Group, 2002). Fatty liver, hyperglycemia, hypoglycemia, essential fatty acid deficiency, hypokalemia, and hyperlipidemia can result when using TPN (UK Cystic Fibrosis Trust Nutrition Working Group, 2002).

3.6. Disordered Eating and Body Esteem

3.6.1. Disordered eating in adolescents.

Disordered eating is increasing among Western adolescents. Not only has the number of dieters increased, the age of onset is decreasing (CPS, 2004). Popular media favor slimness, skewing adolescents' self-perception. Their changing bodies are far from desirable by today's standards, which can lead to disordered eating attitudes and behaviors. Almost half of a group of

publicly schooled teenagers in Ontario, 12 to 18 years old, were dissatisfied with their weight (Jones, Bennett, Olmsted, Lawson, & Rodin, 2001).

Dieting is defined as an intentional, usually temporary, altered eating pattern to try to lose weight (Neumark-Sztainer, Story, Dixon, & Murray, 1998). Cross-sectional data from Canada states that around one in five teenaged girls are on a diet (Jones et al., 2001). Recent studies show that around 60% of teenaged girls, and 25% of teenaged boys, have tried to lose weight in the past (Daee et al., 2002). Among these dieters, there is a high rate of unhealthy dieting behavior which includes using diet pills, fasting, skipping meals, or using very low calorie diets (Daee et al.). While sensible weight loss in the overweight or obese population can have positive health effects, dieting can lead to negative psychological and physiological consequences such as the adoption of more negative health behaviors, a poor body image, decreased self-esteem, loss of electrolytes, minerals, and trace metals, changes in the muscle of the heart, and adverse effects of free fatty acids on the heart muscle (Daee et al.). As well, teens who adopt extreme weight reduction methods (self-induced vomiting and/or use of diet pills) may be at an increased risk for engaging in other high risk health behaviors such as tobacco, alcohol, and drug use, suicide ideation, and unprotected sexual activity (Neumark-Sztainer et al., 1998).

Methods to induce weight loss vary in severity and extremeness. Eight percent of girls (12 to 18 years old) attending public schools in Ontario, reported self-induced vomiting as a method of weight loss (Jones et al., 2001). This is similar to general rates of approximately five to twelve percent of adolescent girls. Other weight loss methods used include fasting, skipping meals, crash diets (22-46% in teenage girls), laxative and diuretic use (one to four percent), use of diet pills (three to ten percent), and smoking (12-18%) (Jones et al.). Another study of 1739

Canadian female adolescents, found that 13% of those between 12 and 14 years of age, and 16% of those between 15 and 18 years of age, had assessment scores (via Eating Attitudes Test-26) reflecting disordered eating habits (Jones et al.). Dieting to lose weight was reported by 23% of the study population; binge eating was reported by 15%; self-induced vomiting was reported by 8.2%; and the use of diet pills was reported by 2.4% (Jones et al.). While females exhibit more disordered eating behaviors, the outcome and course of these disorders are not gender specific (Eliot & Baker, 2001).

Information on dieting and weight loss must be obtained from somewhere. In one study of 62 third to sixth graders in San Francisco, 50% wished to be thinner, with 16% reporting that they had previously tried to loose weight (Schur, Sanders, & Steiner, 2000). The majority of these children received information on dieting from family members (77%), usually a parent, with the media coming in at second place (55%). Schur et al. (2000) feel that parents should be more cautious about how they talk about weight loss and dieting around their children. They must be responsible for educating their children on normal body changes during adolescence, and that adult weight control methods may not be favorable in actively growing children (Schur et al.).

3.6.2. Body esteem in adolescents.

One's self-evaluation of body appearance, of which weight is one aspect, is termed body esteem (Mendelson, Mendelson, & White, 2001). Usually measured by self-report surveys, interviews, or reactions to body-related words, one's feelings about their body can differ from the perceptions and opinions of others (Mendelson et al., 2001). Since females are more likely to hold higher standards for their bodies than males, the role of gender in self-evaluation of appearance is often important (Mendelson et al.). A study of children from Quebec aged 8.5 to

15.4 years, assessed by *The Self-Perception Profile for Children/Adolescents* and *The Revised Body Esteem Scale*, found that girls were more likely to rate their body appearance as negative, when compared to boys, at all ages (Mendelson et al.). These evaluations became increasingly negative as the children aged and/or gained weight (Mendelson, White, & Mendelson, 1996).

A link between self-esteem and body-esteem has been seen in the above study and often, regardless of weight, those who felt good about the way they looked were more likely to evaluate their overall self-worth as being high (Mendelson et al., 1996). There is a lack of research on the role of body esteem and health behaviors, specifically adherence to diet, nutritional supplements, and enzymes, in the CF population. As will be seen in the next section, females with CF are likely to overestimate their body weight (Abbott et al., 2000). This overestimation may lead to unhealthy behaviors in a population whose health is already declining.

3.6.3. Disordered eating and body esteem in CF.

Research on the prevalence of eating disorders in the CF population is sparse. A recent study found that no participants with CF, out of 58, provided self-reported indications of an eating disorder (Raymond et al., 2000). Results from *The Eating Disorder Inventory*, found that control participants were more concerned with being thin than CF participants (Raymond et al.). This could be due to the fact that CF participants tend to weigh less than same aged counterparts. Although they may not be overly concerned with being thin and losing weight, there is a possibility that the opposite reaction may be occurring here- they may be overly concerned with gaining weight, which was not measured using the above tool.

Another study examined children between seven and 12 years of age using *The Children's Body Image Scale* (Truby & Paxton, 2001). This study found that the 76 CF participants and 153

control participants responded similarly to those in the previous study. CF participants had a lower BMI than control participants, despite reporting an artificially high body size (Truby & Paxton, 2001). They also had a lower mean score for body size satisfaction than control participants, while having a higher mean score for body weight satisfaction (Truby & Paxton, 2001). The girls in the study had smaller ideal body sizes than the boys, with 38% of boys desiring a greater body size compared to 19% of girls (Truby & Paxton, 2001). This may give significance to the hypothesis that feelings about weight can be different from feelings about general appearance (Mendelson et al., 2001).

The discrepancy in satisfaction with body size and weight may be explained by the fact that CF participants tend to be shorter than their peers, due to stunting from malnutrition or delayed puberty (UK Cystic Fibrosis Trust Nutrition Working Group, 2002; Pencharz & Morton, 2003; Landon, Rosenfield, Northcraft, & Lewiston, 1980). Caution should be exercised in interpreting the results as some scales, since they may be worded towards a preoccupation with thinness specifically, not general body dissatisfaction.

One's perception of their weight can often differ from their actual weight. Studies indicate that females with CF are more likely to believe that they are overweight, regardless of true body weight (Walters, 2001; Abbott et al., 2000). On the other hand, men with CF generally believe that they are underweight, even if they are at a healthy weight (Walters, 2001). In a study of approximately 1050 participants with CF (median age 23 years), a significantly higher proportion of females with CF who were very underweight (< 85% IBW) or underweight (85-94% Ideal Body Weight (IBW)) perceived themselves as being normal or overweight (29% versus 11% for male participants) (Walters, 2001). Conversely, a higher proportion of the males

in the study who were normal weight perceived themselves to be underweight (41% versus 15% of females). These stats seem to indicate the possible presence of a distorted body image.

When perceived body size was determined for adults with and without CF, males without CF correctly estimated their body size and were content with their current weight, while CF males overestimated their BMI and but desired to gain weight (Abbott et al., 2000). Females without CF underestimated their BMI but desired to lose weight, while CF females were satisfied with their current state, despite underestimating their BMI's (Abbott et al.). This satisfaction may not be seen in teenagers, since they are in a period of growth and adjustment which may be delayed compared to their peers.

The self-perception of one's body weight can impact upon eating behaviors, which can have important consequences in the CF population. Walters (2001) determined that a significantly higher proportion of those who perceived themselves as being underweight were taking oral or enteral supplements (77% versus 30%). Multiple logistic regression showed that a self-perception of being underweight was the strongest predictor of taking such supplements (Walters, 2001). Men who were normal or underweight reported taking a significantly higher dose of enzymes and nutritional supplements versus the women (Walters, 2001). Out of all study participants, 47% reported taking oral or enteral nutritional supplements and 22% reported restricting their fat intake (Walters, 2001). Given the fact that women with CF are more likely to perceive their body as being overweight and that they are less likely to take nutritional supplements, women with CF may be at an increased risk of developing disordered eating behaviors (Walters, 2001).

3.7. Adherence

3.7.1. The notion of adherence.

The words adherence and compliance are often used interchangeably in the literature. Compliance is defined as: "the extent to which patients are obedient and follow the instructions, proscriptions, and prescriptions of health care professionals" (Meichenbaum & Turk, 1987). According to this definition, compliance takes on a passive role of obedience, and non-compliance is often viewed negatively and as being the fault of the patient (Meichenbaum & Turk, 1987).

Adherence, on the other hand, is defined as: "an active, voluntary, collaborative involvement of the patient in a mutually acceptable course of behavior to produce a desired preventative or therapeutic result" (Meichenbaum & Turk, 1987). This definition involves choice on the patient's behalf and when adherence occurs, it is to a "consensually agreed upon plan" (Meichenbaum & Turk, 1987). The switch to the term adherence reflects the work of the patient/client in making decisions about their care and following though with this care (Kettler, 2004). In this study, the word adherence will be used since it gives the patient active, voluntary involvement in their treatment, as opposed to submitting to the demands and requests of others.

Adherence is not static; possibly changing over time, it is a complex behavior and may not necessarily predict similar patterns for all medical regimens and medications (Meichenbaum & Turk, 1987) (see Table 1). Meichenbaum & Turk (1987) divide adherence into a number of behavior related facets:

- Entering into and remaining in a treatment program;
- Attending follow up and referral appointments;
- Taking medication properly;

- Properly following life style changes (ex. diet);
- Properly administering therapies at home; and
- Avoiding health risk behaviors.

Table 1: Different Forms and Causes of Non-Adherence

Type of Adherence	**Causes of Non-Adherence**
Medication	• Failure to fill the prescription • Filling the prescription but failing to take the medication, or only taking part of it • Not following the frequency of dose instructions • Taking medication that was not prescribed
Treatment	• Delay in seeking care • Failure to enter treatment programs • Not keeping appointments • Premature termination
Behavior	• Not taking recommended preventative measures • Incomplete implementation of instructions • Sabotaging of treatment regimen • Nonparticipation in prescribed health programs • Creating one's own treatment regimens to fill the gaps of what one believes one's health care provider is overlooking • Substituting one's own program for recommended treatment regimens

Source: Meichenbaum & Turk, 1987.

General rates of adherence are in the vicinity of 30 to 60%, taking into consideration varying levels of non-adherence such as (a) never adhering to any recommendations, (b) adhering to some but not all, (c) initially adhering but then non-adhering after some time, and (d) adhering but not in the appropriate way (Masek, as cited in Meichenbaum & Turk, 1987). Adherence often decreases for prophylactic agents, to an average of 30 to 35% (Marston, as cited in Meichenbaum & Turk, 1987).

Part of the challenge in determining rates is due to the difficulty in accurately measuring

adherence. Health care providers may assume that their clients are adherent when they really are not, they may not inquire about levels of adherence, and they may be the last to know about such non-adherence due to the client's withholding of information (Meichenbaum & Turk, 1987). Adherence levels are difficult to gauge for other reasons such as (a) adherence may be viewed in different ways, (b) adherence may be defined in different ways, (c) adherence often changes from situation to situation, and (d) adherence can be measured in various ways (Meichenbaum & Turk, 1987). Self-reports may be subject to inaccuracies, manipulation, and bias (Cluss & Epstein, 1985). Over-reporting may occur if the participant desires to be viewed in a positive manner, and self-monitoring progress may alter behavior so it no longer reflects usual adherence patterns (Meichenbaum & Turk, 1987). However, while imperfect, these measurements are all that are available at this point in time.

Measurements of adherence can vary according to the definition of adherence. Sometimes the percentage of adherence is determined according to a formula, [(number of doses taken/ number of doses prescribed) x 100], whereas sometimes it is determined according to a pre-determined standard (such as 80% or more of recommendations being considered adherent) (Cluss & Epstein, 1985). Since this may lead to over- or under-estimations of adherent individuals, adherence should be kept to the minimally accepted level of a medication which will produce the desired benefit (Cluss & Epstein, 1985).

Levels of adherence can vary from time to time or from treatment to treatment. Lask (1994) divides non-adherence into different levels- fully or almost fully adherent, partially adherent, and predominantly adherent. Patients can be considered adherent if they claim to be so, and do not show discrepancies between self-measured and laboratory-measured criteria. Those who openly

admit to a certain level of non-adherence, with a marked discrepancy between the self-measured and laboratory measured criteria, are primarily considered non-adherent (Lask, 1994). Below is a list of various methods that can be implemented to measure adherence:

- Interview;
- Behavioral measures;
- Self-report;
- Clinical rating;
- Self-monitoring;
- Biochemical indicators;
- Pill counts of unused tablets;
- Record of broken appointments;
- Tallies of refills of medications;
- Marked-sign techniques (inactive or false markers embedded in treatment package); and
- Clinical outcome improvement or stability in medical condition or symptoms (Meichenbaum & Turk, 1987).

What determines whether one is adherent or not? Garcia, Schooley, and Badaro (2003) refer to an "adherence trilogy" which is composed of three main facets. The first facet is information- in order to adhere to a treatment one must first understand the disease/illness, understand what the implications are for their lives, and be aware of the consequences of treatment non-adherence. Motivation is the second facet of the trilogy-patients must be motivated in order to adhere (Garcia et al., 2003). It is not simply enough to know the information; one must want to apply the facts to his/her life. Factors such as depression may negatively impact adherence levels (Horenstein, Moustier, & Martineau; as cited in Garcia et al.). Behavioral skills are the last facet of the trilogy (Garcia et al.). While it is virtually effortless for a doctor to prescribe a treatment or medication, it will have to be adopted into the person's life. Treatments or drugs may lead to an alteration of a person's habits and lifestyle and, in some cases, the two may be incompatible (Garcia et al.).

3.7.2. Adherence and CF.

Non-adherence, a common problem in the CF population, is an important issue in the management of CF. Reasons for non-adherence in other clinical populations are often transferable to those with CF, which takes into consideration the history and demands of the disease (Koocher, McGrath, & Gudas, 1990). Cluss & Epstein (1985) indicate that research has found adherence to be lower among those with chronic diseases or who use medication prophylactically. Unfortunately, CF falls into both of these categories.

Problems with adherence in CF were reported as early as 1981, when a study of 58 CF participants found that 90% adhered to medication, 40% to chest physiotherapy, and only 20% to dietary treatments (Passero, Remor, & Salomon, 1981). Ten years later, a study looked at the perceived adherence to medication and diet as reported by those with CF, a parent, and physician. Mean perceived adherence to medication was around 75%, and that of diet was between 50 and 75% (Gudas, Koocher, & Wypij, 1991). Reported perceived adherence levels for diet resulted in a large degree of confusion and inconsistency between the parent, physician, and self (Gudas et al., 1991). Such a discrepancy of perceived adherence between child and parent was also found in a study by Ievers et al. (1999). Here, self-reported adherence to airway clearance treatments was 90% and parent-reported adherence was 85%, adherence to aerosol medication was 63% and 64%, respectively, and adherence to pancreatic enzymes was 70% and 68% (Ievers et al.).

Adherence to lifestyle changes has been found to be lower than adherence to medication. A study of 96 participants with CF (nine to 16 years old) found that the average rating for adherence to medication (including enzymes) was "usually followed recommendations", but that

for dietary recommendations averaged "rarely followed recommendations" to "followed recommendations 50% of the time" ((DeLambo, Ivers-Landis, Drotar, & Quittner, 2004). Reports from males showed that they followed dietary recommendations better than females, which was mirrored in the mother's responses where gender was responsible for 6.1% of the variance in reported adherence (DeLambo et al., 2004).

Among the list of reasons for non-adherence, teenaged female diabetics have been shown to alter their level of adherence to insulin as a method of weight control (Bryden et al., 1999). Thirty percent of diabetic females and 45% of diabetic females with microvascular complications, misused insulin to control their weight, as opposed to none of the males (Bryden et al.). A Canadian study stated that between 11% and 34% of teenagers and young adults with diabetes have misused insulin to reduce or control weight (Jones et al., 2001). While no studies were found showing evidence of the misuse of pancreatic supplements, this mechanism could provide a simple weight-control method for those with CF. However, non-adherence to enzymes or diet, possibly resulting in unhealthy eating habits, can impair the health of those with CF, leading to a poor nutritional status and thus a poorer prognosis (Pfeffer, Pfeffer, & Hodson, 2003).

3.8. Factors that Affect Adherence

While it is important that the CF population adhere to medical and dietary recommendations, various factors may lead to non-adherence. While it is assumed that adherence will result in a favorable clinical outcome, there are many other factors which may play a role (Meichenbaum & Turk, 1987). Two such factors occur if the effect of adherence is statistically independent of the

medication/treatment, and if the medication/treatment has poor efficacy, in which case it would be misleading to attribute a negative outcome to non-adherence (Cluss & Epstein, 1985). As is the case for those with CF, even with diligent adherence health is expected to deteriorate with age, which may give the impression of non-adherence or simply frustrate the adherent individual. With CF, adherence offers the hope of a prolonged period before decline, but not a cure.

3.8.1. Medical demands.

Medical and treatment demands influence adherence, with highest rates being seen for treatments which involve the administration of medications, high supervision and monitoring, and an acute onset of the medical condition (Meichenbaum & Turk, 1987). Adherence is often negatively correlated with the duration of the treatment, the number of medications or treatments being taken at one time, and related side effects (Cluss & Epstein, 1985; Garcia et al., 2003; Lask, as cited in Kettler, Sawyer, Winefield, & Greville, 2002). Unfortunately, treatment for CF often involves intensive daily routines involving chest physiotherapy up to four times a day, the time consuming administration of medication by nebulizer, taking up to 80-100 pills a day, and eating large quantities of high-calorie foods at three meals and three snacks (Gudas et al., 1991; Kettler et al., 2002). Daily adherence to, and remembrance of, such a complex and time consuming regimen is challenging and may lead to an increased risk for non-adherence (Kettler et al.; Conway, Pond, Hamnett, & Watson, 1996; Abbott, Dodd, Bilton, & Webb, 1994). In addition, non-adherence may also occur when the realities of CF's demands do not coincide with the goal of a "normal" life (Conway et al., 1996).

As part of the individual's life and care, the medical team often plays a significant role. Relationships are often developed over many years (Kettler et al., 2002), and it has been shown

that trust in the team is an important predictor of adherence (Lask, as cited in Kettler et al.). Adherence is also predicted by the self-sufficiency of the individual, by playing an active role in his/her doctor's appointments (Litt, Cuskey, & Rudd, 1980). It has been shown that those who attend regular clinics have been found to be more adherent (Conway et al., 1996). Below is a list of some medication related factors which contribute to non-adherence:

- Adverse real or imagined effects;
- Complex regimens including multiple drugs or frequent dosing;
- Unpleasant administration including injections or suppositories;
- Unpleasant taste or smell;
- Drugs that are similar in appearance or with similar names; and
- Inconvenient or restrictive precautions such as the exclusion of alcohol, foodstuffs such as cheese, and specific requirements related to administration in relation to meals (Garcia et al., 2003).

In addition, below is a list of some patient related factors contributing to non-adherence:

- Misunderstanding of prescribing instructions;
- Frequent changes to drug regimens;
- Multiple physicians or health care providers prescribing medication;
- Limited faith in the effectiveness of the medication or in the care provider;
- Inability to read written instructions;
- Forgetfulness or confusion;
- Denial of the illness or its significance;
- Anger about the illness;

- Apathy;
- Depression;
- High stress;
- Reduction, disappearance, or fluctuation in disease symptoms;
- Concern about taking drugs, including fear of becoming addicted;
- Inability to afford medication;
- Physical difficulties limiting access to or use of medication such as: problems swallowing tablets, difficulty in opening drug containers, difficulty in handling small tablets, and difficulty in getting prescriptions filled;
- Inability to distinguish colors or identifying markings on medications;
- A history of non-adherence to regimens in the past;
- Problems with required assistance in the home;
- Concurrent substance abuse;
- Limited education about the illness or the need for medication; and
- Limited social or family support (Garcia et al., 2003).

3.8.2. Gender and age factors.

Generally, poor adherence is often related to adulthood, with female non-adherence being 35% higher than male non-adherence (Lask, 1994). Children are often forced to adhere to medical treatments by their caregivers and as the child ages his/her health is no longer the responsibility of the parent/guardian (Pfeffer et al., 2003). This encouraged autonomy may lead to poor health choices, possibly caused by rebellion (Landau, 1995).

Growing into adolescence brings unique challenges such as peer pressure and the need to fit

in. Certain aspects of CF are quite noticeable such as a constant cough, and taking medication. Peers may exert pressure on CF adolescents to stop taking prescribed medication, such as enzymes, and to look and/or behave according to an accepted norm (Conway et al., 1996). Also, CF adolescents may feel embarrassed to take enzymes in front of their friends (Conway et al.).

In addition to this pressure, studies have shown that adolescents and young adults with CF may suffer from different emotional problems and/or psychosocial resistance such as: low self-esteem, depression, denial of illness, an internal struggle for control or autonomy, social dependency, distorted body image due to an altered physical appearance, a denial of sexuality often with a latent initiation of intimate relationships, isolation, and the awareness of the future and death (Landon et al., 1980; Simmons et al.,1985). Being consumed with thoughts of death, which may be unrealistic, can lead to detrimental behaviors and non-adherence (Johannesson, Carlson, Brucefors, & Hjelte, 1998). A distorted body image may lead to low body esteem, and may affect supplement use and/or adherence to diet. Since CF females are more likely to overestimate their body weight, as described in section 4.5.2., gender may play an interesting role with regards to body esteem and adherence.

3.8.3. Behavioral and psychosocial factors..

Adherence rates associated with a chronic medical condition, such as CF, are often lower than rates for an acute condition (Meichenbaum & Turk, 1987). A person's behavior and psychosocial equilibrium (or lack thereof) may play a role in adherence. In terms of behavior and adherence, individuals can be refusers, procrastinators, or deniers (Lask, 1994). Refusers will not hide the fact that they do not adhere to recommendations, stating they do not feel the treatment is necessary, they feel the treatment is worse than the symptoms, they feel that the treatment is

pointless, or that they will not be told what to do or be controlled by medical staff (Lask, 1994). Procrastinators are more resistant to admit to non-adherence, but will often confess to occasional non-adherence, stating that they will adhere in the future-which may or may not occur (Lask, 1994). Deniers will not admit that they do not follow medical recommendations, even though this is probably the case (Lask, 1994).

Koocher et al. (1990), give examples of how psychosocial factors may lead one with CF to resist adherence. Such examples include: control struggles between parents and other authority figures, cultural and peer pressures, the drive to have a 'normal' life, denial, avoidance, and living in a chaotic home environment (Koocher et al.). See Case Scenario 1 for an example of a form of psychosocial related resistance.

Case Scenario 1: Cultural Pressures

Situation: Jose believed that one ought not to go to the doctor's office "unless you're really sick". Neither he nor his family believed that the preventative therapies prescribed by his physician really helped his CF very much. Besides, he was a "man" of 16, and wanted to hold onto his tough macho image among the males in his peer group. His family placed little or no emphasis on his adherence to the prescribed treatment regimen, although his grandmother often brought over herbal teas and other folk remedies which Jose dutifully imbibed.

Commentary: The meaning of illness in general and CF in particular varies widely across social roles and cultural groups. When one's social context does not support acknowledging and treating a chronic medical condition, denial takes over. The result can be a lack of preventative care and delay in seeking treatment for exacerbations of the illness. Jose is clearly able to participate in the treatment regimen approved by his social context (i.e., herbal teas), but is less willing to accept the more alien medical advice. At the same time, he probably does not openly reject his physician's advice when they talk; leading to the misconception of the part of the physician that Jose is following prescribed treatment.

Source: Koocher et al., 1990

3.8.4. Perception of severity and self-care.

Two factors which may play a role in adherence, which are also key factors of *The Health Beliefs Model* which will be discussed in the Theoretical Framework section, are the perceived severity of the disease and the patient's perceived self-care. It has been shown that physicians perceive disease severity and self-care differently than do patients (Abbott, Dodd, & Webb, 1996). When 60 CF patients, mean age 21 years, and their physician and close companions (partner or mother) were questioned about disease severity, significant discrepancies were found (Abbott, Dodd, & Webb, 1995). Each was asked to rate disease severity on a five point Likert scale ranging from below average (rating 1), to well above average (rating 5). Ratings were similar between the participants and their close companions (4.05 ± 1.10 and 4.20 ± 0.79, respectively); however, there was a significant difference between the ratings of the participants and their physicians (2.92 ± 1.15) (Abbott et al., 1995).

While physician ratings were positively correlated with FEV_1 scores, WFH, and the Shwachman score, participants with the most severe disease viewed themselves as being above or well above average (rating of 4 and 5, respectively) (Abbott et al., 1995). Eight of the participants, having been assessed for heart-lung transplants, were rated in the "poor" (rating 1) or "below average" (rating 2) category by physicians, but were rated in the "well above average" or "above average" category by themselves and their close companions (Abbott et al.). This suggests that those with CF may overestimate the quality of their health. Those who underestimate the severity of their illness may be more likely to be non-adherent than those who do not (Czajkowski & Koocher, as cited in Abbott et al.).

Self-care was also considered in this study. Participants, their doctors, and their companions

were asked to rate the quality of the self care of the individual with CF on a six point Likert scale, ranging from" very poor" (rating 1) to "excellent" (rating 6). Self-care was defined as the overall self-management of the disease, including physiotherapy, exercise, and taking medications (including enzymes and vitamins). Ratings were significantly different between the self reports (4.30 ± 1.10), physician reports (3.37 ± 1.23), and companion reports (5.02 ± 0.66) (Abbott et al., 1995). Eighty percent of the companions had ratings in the categories "very good" (rating 5) or "excellent" (rating 6) (Abbott et al.). However, only 15% were rated at this level by their physicians (Abbott et al.). This suggests that physicians may not have an accurate picture of such levels.

A factor which plays a role in self-care is the perceived personal control over disease progression, also known as locus of control (Abbott et al., 1996). An internal locus of control is one in which an individual feels that they can determine, or play a role in, the outcome of their disease. The chance locus of control exists when an individual feels their health is determined by chance, luck, or fate (Abbott et al.). Lastly, the powerful others locus of control places other people such as doctors, nurses, or family members, in the pivotal role of influence on personal health outcomes (Abbott et al.).

The locus of control theory assumes that people will take control of their disease management, and adhere to treatment, if they exhibit an internal locus of control (Abbott et al., 1996). However, this may not always be the case as was discovered in a study of 60 participants with CF (mean age 20.98 years) which indicated the type of locus was similar regardless of reported adherence to enzymes and vitamins (Abbott et al.). Those who were adherent to enzymes (rating of 'I always take my enzymes as prescribed' on *The Manchester Adult CF*

Compliance Questionnaire) scored higher on the chance scale than those in the other adherence categories. Those who were adherent or partially adherent (rating of 'I always take my enzymes as prescribed' and 'I usually take enzymes as prescribed') felt that their disease outcome was greatly under the influence of powerful others, when compared to those who were non-adherent (Abbott et al.). With regards to vitamin adherence, those who were adherent had a strong belief that their health was affected by chance and powerful others, when compared to those who were partially adherent or non-adherent (Abbott et al.).

3.8.5. The role of knowledge.

Parental and self knowledge of one's medical treatment can greatly affect the rate of adherence (Ievers et al., 1999; Koocher et al., 1990). Medical information is often intricate and can be hard to understand when combined with a complex diagnosis. Information needs to be provided at a developmentally appropriate level, and updated throughout the life cycle (Koocher et al.).

Ievers et al. (1999) studied 45 mothers and their CF-children (median age was eight years) to determine if any relationship was present between knowledge and adherence. Mothers reported all known prescribed medications/treatments that their child was asked to take. While reports of the dose of enzymes taken with meals and snacks were significantly related to medical charts, 11.5% did not know the exact maximum dosage which was prescribed with meals (Ievers et al.).

When looking at child-reported meal enzyme dosage, there was a significant association between the reported amounts and the prescribed dose, but 16.7% could not give the greatest amount that was prescribed, and 12.5% stated that they should take two to four pills less than was prescribed (Ievers et al., 1999). When looking at enzymes taken with snacks, there was a

moderate association between the child-reported numbers and those prescribed, but 42.9% could not give the highest dose prescribed (Ievers et al.). There was also a significant association between accuracy and age, with regards to enzymes taken with snacks, but there was no gender difference (Ievers et al.). After individual differences were controlled for, child and parental knowledge (especially maternal) about CF and the treatment regimen, accounted for a significant part of the differences in the child's reported adherence (Ievers et al.).

In addition to inadequate knowledge, Koocher et al. (1990) state that educated non-adherence can also occur. In this case, non-adherence occurs despite having received adequate education. The individual is aware of the reasons why medical treatments are prescribed and what will happen if they do not follow the treatments (Koocher et al.). Despite knowledge of the medical treatment, non-adherence may occur because of: disagreements with their clinicians (i.e., holding different personal and health values and/or interpreting their health in a different way; see Case Scenario 2) (Wright & Morgan, 1990), unwanted side effects (Smith, Rosen, Trueworthy, & Lowman, 1979), having to take a bad tasting treatment (nutritionally, this can apply to supplements) (Conway et al., 1996), coming to a personal decision to give up (feels nothing makes a difference to health; see Case Scenario 3) (Koocher et al.), and a deciding to "trade" certain treatment aspects for a perceived higher quality of life (Koocher et al.).

Case Scenario 2: Mixed Signals

Situation: Judy, age 18, moved across the country to attend college. In doing so, she shifted her medical care from one CF clinic to another. Her new physician suggested she should consider substituting aerobic exercise for some chest physical therapy, and altered some of her prescriptions without providing a detailed explanation for the changes. When she tentatively mentioned how strongly her former caregivers had insisted on her old regimen, she was simply told "we've found these changes work well". Judy felt confused and torn between her positive feelings for the advice of her long-time care providers (to whom she attributed her relatively good state of health) and the unexplained changes proposed by a stranger on whom she was now to rely for medical care. The result was a half-hearted application of the new advice as she attempted to maintain her former regimen, and misled the new physician as to her actual behavior.

Commentary: There are many controversies regarding the efficacy of some aspects of CF treatment among physicians who are expert in treating this illness. When patients develop a consistent routine which "works" for them, they are reluctant to change without clear rationales. In addition, what is "clear" to the physician may be opaque to the patient. When a change of caregivers occurs, a patient's need to adhere to the treatments prescribed may be increased, especially when the old routines are associated with former caregivers who are highly regarded or emotionally significant.

Source: Koocher et al., 1990.

Case Scenario 3: Palliative Adherence

Situation: John, 29, had severe lung disease, mild congestive heart failure, and diabetes as complications of his CF. Although he tried to keep his strength up by eating as much as he was able, he often found himself vomiting after chest physical therapy (CPT). The CPT sessions often left him feeling especially weary, as well. Although he fully understood the nature of his illness and the rationale for treatment, John decided that CPT was simply not worthwhile, any longer. He knew that he was in the terminal stages of his illness, and made a reasoned choice not to continue his CPT.

Commentary: Choosing to cease a treatment in the face of a terminal illness is not a new issue or one that is unique to CF. The patient may well determine that the benefits of the regimen are far outweighed by its costs, long before the medical team reaches the conclusions. This can become a source of friction and anger, should control struggles develop over implementing a medical regimen. At the same time, it is not unusual for a patient to be ambivalent regarding the weight of the costs and benefits, especially as symptoms wax and wane.

Source: Koocher et al., 1990.

Those who do not automatically listen to orders from health professionals may be labeled "problem patients", since some health professionals may expect to hold the power in the relationship, and see longevity as the number one priority for health care. However, this may not be the case with those who have a chronic illness, and the patient's need to hold more autonomy with regards to their own health care should not be viewed as negative (Wright & Morgan, 1990).Koocher et al. (1990) conclude that "sometimes, it is more important to assist patients in getting the information needed to make informed choices that reflect their particular needs, values, and goals, than to focus solely on adherence. This might mean that some patients receive less then optimal medical care…..while gaining a degree of quality of life that is more important to them."

3.8.6. The role of responsibility.

As the child ages, he/she is given more responsibility over his/her medical treatment. Drotar

and Ievers (1994), in a study of 26 mothers of CF children and adolescents (ages four-14 years), found that as the age of the child increases, they are given more responsibility over their health care. The percentage of responsibilities that were shared among parents and children increased between four and seven years of age (19%) and eight to ten years (32%), but remained about the same from 11 to 14 years (34%) (Drotar & Ievers, 1994).

It is important for children to have the knowledge base that will allow them to be responsible for their health. If children are given responsibility before they are mature, detrimental effects to their health may occur (Drotar & Ievers, 1994). It is important for parents and physicians to gauge the child's level of information about CF and his/her treatment, before various levels of independence are given to the child (Drotar & Ievers, 1994). As suggested by Ievers et al. (1999), "children with CF do not necessarily become more knowledgeable about their prescribed treatment-related tasks throughout the elementary school years".

3.8.7. Positive predictors of adherence.

Certain factors are positively correlated with adherence. These include: the age of the individual (younger patients are more likely to adhere) (Gudas et al., 1991), a high degree of optimism (Gudas et al.), a higher degree of child knowledge about CF and its treatments (Gudas et al.), level of education, a high socio-economic status (Gudas et al.), and family expressiveness (Patterson, as cited in Lask, 1994). Those who tend to worry are more likely to adhere to virtually all aspects of treatment, including enzyme and vitamin therapy (Abbott et al., 1994, 1996). While theory suggests that those with an internal control belief would be more likely to adhere than those who believe health outcomes depend upon chance or other people, Abbott et al. (1996) discovered that those with little self-control over the course of their disease had greater

reported adherence levels than those with an external control belief.

3.9. Summary

Cystic fibrosis is a multidimensional, lifelong illness, not only affecting the physiological aspects of one's body but emotional and mental aspects as well. CF related malnutrition can lead to a low BMI, vitamin and mineral deficiencies, a short stature, delayed puberty, a decrease in lung quality, and even osteopenia or osteoporosis.

CF adolescents may have a distorted body perception and low self-esteem due to factors related to malnutrition such as a short stature and delayed puberty. Canadian adolescents, particularly females, frequently display disordered eating behaviors (CPS, 2004). This can also occur in CF adolescents who are not satisfied with the way their bodies look. However, in this case, disordered eating behaviors may take the form of wishing to remain slim and thus restricting caloric intake which should be 120-150% of recommendations, or not adhering to enzyme therapy. This restriction may have detrimental effects on their long term health.

CF-created slimness may be perceived as ideal, in a society that is intent on weight-loss, leading some to compromise health for slimness, by not adhering to enzymes and medical/dietary advice. Clinicians and dietitians must be aware of the dynamic problems faced by those with CF as they age. Adolescence brings with it confusion and emotional fluctuations for everyone; however, this period of adjustment is further complicated by the existence of a chronic condition.

Section 4: Theoretical Framework

4.1. Theory of Planned Behavior

In 1980 Ajzen, and later Ajzen and Fishbein, developed *The Theory of Reasoned Action*, after attempts were made at estimating the discrepancy between people's attitudes and behaviors (Universiteit Twente, 2004b; Fishbein & Ajzen, 2005). The early premise of the theory assumed that performing a certain behavior was voluntary, which was not the case at all times. The theory was then adapted to include the parameter of perceived behavioral control and was re-named *The Theory of Planned Behavior* (TPB) (Universiteit Twente, 2004b).

The main assumption of TPB is that an individual's behavior is determined by their intention to perform the behavior. Intention is defined as "the cognitive representation of a person's readiness to perform a given behavior, and it is considered to be the immediate antecedent of behavior" (Universiteit Twente, 2004b). Intention is shaped by three things:

I. The individual's attitude toward the behavior in question;

II. How the individual believes people will view the behavior (subjective norms); and

III. The individual's perceived behavioral control over, and ability to perform, the behavior.

Therefore, if one believes they have a strong sense of control over a certain behavior, their intention to perform the behavior should be greater (Universiteit Twente, 2004b).

The issue of independence can play a duel role, acting as an enabler or a barrier. The lives of teenagers are often tightly controlled by others such as parents/guardians, and the education system. Teenagers, while being offered more freedom as they age, are still bound by various rules and regulations which they have very little control over. In fact, the area of personal health may be one of the few that are primarily under their control. Increased independence and

autonomy can lead to an abuse of power on their part (leading to non-adherence), but it could alternatively lead to the valuing of independence and informed, supported decision making, fostering adherence.

4.2. The Health Belief Model

The Health Belief Model (HBM), developed first in the 1950's by social psychologists Hochbaum, Rosenstock, and Kegels, is a psychological model that tries to explain and predict health behaviors (Universiteit Twente, 2004a). Similar to the TPB, the HBM assumes that people will take a certain health related action if they feel a negative health condition can be avoided, and if they believe they can successfully take the action. There are six key terms central to the HBM:

I. Perceived Susceptibility: Perceived chances of getting a condition;
II. Perceived Severity: Self-opinion of how serious a condition is, including possible consequences;
III. Perceived Benefits: The belief (or non-belief) in the efficacy of the action to lessen the risk, or seriousness, of impact;
IV. Perceived Barriers: The perceived tangible and psychological costs of the action;
V. Cues to Action: Strategies present to activate "readiness" (education, symptoms, media information); and
VI. Self-Efficacy: Self-confidence in the ability to take the action (Universiteit Twente, 2004a).

As well, the theory makes assumptions about the following three things, as noted by Cluss & Epstein (1985):

1) The individual's personal readiness to take part in preventative health behaviors;
2) The individual's evaluation of the likelihood and probable efficacy of taking part in the behavior; and
3) The cues to action which must take place to induce health-related behaviors.

With regards to term I, perceived susceptibility, the theory suggests that if one perceives they are susceptible to poor health, they may be more likely to adhere (Abbott et al., 1996). While anyone with CF realizes they are susceptible to the disease, the degree of perceived susceptibility to its complications may vary. Teenagers may suffer from an egocentric belief in which no one and nothing can harm them, leading to increased risk-taking behavior (Brown, 2002). It has been shown that those with CF may exhibit this belief, although not to the same extent as the general population (Nasr, as cited in Yi et al., 2003). Adolescents may not have the cognitive ability to comprehend long-term consequences of actions and, therefore, may be unable to judge potential future negative health effects caused by non-adherence (Brown, 2002).

For key term II, perceived severity, the HBM suggests that there should be a positive relationship between the perceived severity and adherence. However, this has been refuted in a study of CF participants which concluded that in cases of a life threatening disease, the relationship between perceived severity and adherence may be more complicated than the theory postulates (Abbott et al., 1996). Here, adherence followed a U shaped curve, being lowest when severity was seen as either low or high, and highest when severity was seen as moderate (Abbott et al., 1996). People who perceive that they have a mild disease may feel they do not need the prescribed treatments; whereas, people who perceive they have a severe disease may be unwell or unable to manage their treatments, or feel they receive little benefit from them (Kettler, 2004).

In addition, as described in Section 3.8.4., it has been suggested that the CF population may overestimate the quality of their health (Abbott et al., 1995). Those who underestimate the severity of their illness are more likely to be non-adherent than those who do not (Czajkowski & Koocher, as cited in Abbott et al., 1995). Overall, for the CF population, a more fair assumption may be that if the perceived severity becomes great, people may avoid perceptions and behaviors which reinforce the gravity of the situation (Abbott et al., 1995).

Key term III is the perceived benefits of taking an action, and will be discussed in Section 7.6. Key terms IV and V in *The Health Belief Model* are the perceived barriers to an action, and self-efficacy in performing an action, which mirror factors in the TPB. Cues to action, such as health communication and education by health professionals, can be key enabling factors, as was seen in the study on child and parental knowledge of the child's medical treatment (Ievers et al., 1999). CF clinics are multi-disciplinary and involve the expertise of such professionals as doctors, nurses, dietitians, physiotherapists, and social workers/psychologists (CCFF, 2003). Working together, such professionals can give patients the resources needed to enable them to make informed decisions that will be beneficial to health. Self-care, described in more detail in Section 3.8.4., may be overestimated in the CF population, leading to a distorted picture of adherence when it is not as high as it could be (Abbott et al., 1995).

4.3. Summary

In conclusion, the *Theory of Planned Behavior* and the *Health Belief Model* are two theoretical frameworks which can be applied to this study. While specific questions addressed by the theories are not directly asked in the study, indirect evidence of each will be seen when rates of adherence are determined.

Section 5: Methodology

5.1. Research Design

This study was conducted via mailed out surveys to participants as described in the following section.

5.2. Participants

5.2.1. Population of interest.

The population of interest for this study was adolescents in Atlantic Canada, aged 13 to 20 inclusive, who have CF and fit the inclusion criteria as listed in Section 5.2.2. Due to resource limitations, the sampling frame was chosen to encompass pediatric CF clinics in Atlantic Canada. Clinics at *The IWK Health Centre* (IWK) in Halifax, Nova Scotia (NS) and *The Janeway Children's Health and Rehabilitation Centre* (Janeway) in St. John's, Newfoundland (NL) were chosen by convenience cluster sampling, since these are the only clinics in the area. The scope of the IWK clinic extends to the provinces of Nova Scotia, New Brunswick, and Prince Edward Island. Therefore, all four Atlantic Provinces were taken into consideration.

5.2.2. Inclusion and exclusion criteria.

Participants were included in the study if they had CF and were between 13 and 20 years old, inclusive, from the initiation date of the recruitment to the end of the 2006 calendar year. Participants could be male or female and must have attended a pediatric clinic at the IWK or Janeway. Female participants could be post or pre-menarche, which was not assessed. Participants must have been pancreatic insufficient and prescribed pancreatic enzymes by a physician.

Participants were not excluded due to current nutritional status and/or body mass index, as the study proposed to investigate the participant's nutritional status. Psychological health status did not prevent inclusion as well, as body-esteem was explored in the study.

Participants with CF who also had any of the following were not included in the study due to confounding factors: diabetes, a diagnosed eating disorder, or those who were pregnant or lactating. This is due to the possibility of altered eating behavior due to the presence of an additional medical condition.

5.2.3. Recruitment.

Patients attending the clinics in Halifax, NS and St. John's, NL, who fit the inclusion criteria were invited to participate in the study. A designate from the clinics, the nurse coordinator at the Janeway and the dietitian at the IWK, were responsible for addressing and mailing the packages to eligible participants. Since clinic appointments typically occur once every three or four months, it would have been difficult to recruit participants in a timely fashion if the packages were handed out by the designate and explained in person. Face-to-face contact with participants and the principal investigator was not feasible due to the concerns of bacterial transmission in this population.

The information package given to each potential participant contained:

- Study *Cover Letter* (Appendix A and B) which introduced the principle investigator and the study;
- *Instructions* (Appendix C) for completing the questionnaires;
- One copy of the study *Debriefing Form* (Appendix D and E) detailing the nature and purpose of the study, the requirements of participation, and the rights of each participant;
- The following questionnaires/forms:

- The *Three-Day Food Diary* (Appendix F);
- *The Eating Attitudes Test-26* (Appendix G);
- *The Body Esteem Scale for Adolescents and Adults* (Appendix H); and
- *The Adherence Survey* (Appendix I).

- The *Communication of Results Card* (IWK) or the *Draw Card* (Janeway) which offered participants a chance to obtain a copy of the completed results and/or to be included in a draw for one of two Empire Theatres ™ Gift Certificates (Appendix J); and
- One self-addressed stamped envelope, for return.

Potential participants were asked to read the *Debriefing Form* with a parent/guardian to help them decide if they should participate in the study. This was stated in the study *Cover Letter*. Packages were mailed to potential participants between November 2005 and January 2006.

5.2.4. Consent protocol.

As per the rules and regulations for ethics approval, studies that are done at *The IWK Health Centre* and *The Janeway Children's Health and Rehabilitation Centre* via mailed out surveys do not need to obtain written consent. Consent is assumed upon return of the forms.

5.3. Data Analysis

5.3.1. Nutritional intake.

Participants were requested to complete a *Three-Day Food Diary* to assess their nutritional intake. To analyze food diaries, the USDA Nutrient Database for Windows, HealtheTech Search SR-18, was used (USDA Nutrient Data Products and Services, 2006). When items were not listed in the database, they were either found via the manufacturers' website and/or item

packaging. Examples of websites consulted for nutritional information are: www.greco.ca, www.becel.ca, www.pepsi.ca, and www.generalmills.ca. Examples of packaging which was referred to for nutritional information are: Kraft Dinner, Kellogg's Nutrigrain Bars, Mr. Noodles, and General Mills Dunkaroos.

5.3.2. Estimated Energy Requirement.

While energy amounts in the literature have been recorded in terms of Recommended Dietary Allowance and Recommended Daily Allowance (Roy, Darling, & Webber, as cited in Pencharz & Durie, 1993; Creveling & Gardner, 1997), the introduction of the Dietary Reference Intakes (DRI) resulted in a change to the method of estimating one's daily caloric needs. According to Health Canada, the Estimated Energy Requirement (EER) is used to determine individual caloric requirements. Participants were asked to self-report their weight and height on the AS in order to determine their EER's, %IBW, and BMI_p. To determine each participant's EER, the following formulas, for use in children and adolescents from nine to 18 years old, were used (Health Canada, 2005b):

a) Females: EER (Cal/day) = 135.3 – 30.8 x age + PAC* x [26.7 x wt (kg) + 903 x ht (m)] + 25; and

b) Males: EER (Cal/day) = 88.5 – 61.9 x age + PAC x [26.7 x wt (kg) + 903 x ht (m)] + 25.

*PAC refers to Physical Activity Coefficients. See Table 2 for PAC values. Below is a description of each physical activity category (Health Canada, 2005b).

a) Sedentary: Engages in typical daily activities such as household tasks, and walking to the bus.

b) Low Active: Engages in typical daily activities + 30-60 minutes of daily moderate

activity (example: walking at five to seven km an hour).

c) Active: Engages in typical daily activities + at least 60 minutes of daily moderate activity.

d) Very Active: Engages in typical daily activities + at least 60 minutes of daily moderate activities, + an additional 60 minutes of vigorous activities or an additional 120 minutes of moderate activities.

Table 2: Physical Activity Coefficient (PAC) Values According to Physical Activity Category

Gender (3-18 years old)	**Sedentary PAC**	**Low Active PAC**	**Active PAC**	**Very Active PAC**
Males	1.00	1.13	1.26	1.42
Females	1.00	1.16	1.31	1.56

Source: Health Canada, 2005b.

5.3.3. Nutritional status.

There are various methods of assessing the growth, and indirectly nutritional status, of those with CF. The executive summary from *Dietitians of Canada* (DC), *CPS, The College of Family Physicians of Canada* (CFPC), and *The Community Health Nurses Association of Canada* (CHNA) (2004), recommends the use of the CDC's BMI- for-age charts to calculate BMI percent. It also recommends the use of more traditional methods of assessment, including the %IBW.

A study conducted in 2004 on 13021 participants with CF aged two to 20 years, looked at the difference between the % IBW and BMI_p values when assessing malnutrition in children with CF (Zhang & Lai, 2004). Results indicated that similar weight estimates were found using both methods for children who were of average height (between the 25^{th} and 75^{th} percentiles) (Zhang & Lai, 2004).

The % IBW and BMI_p differ in that they: use different units (percentage relative to the ideal value and percentile ranking, respectively), have different scales (100% being optimal for % IBW, and 50^{th} percentile being optimal for the BMI_p), and have different underlying assumptions (Zhang & Lai, 2004). The % IBW assumes that the ideal weight for a given height is at the same percentile ranking as the height-for-age (Zhang & Lai, 2004). The BMI_p assumes that the ideal weight for a given height is equal to the 50^{th} percentile value for that age (Zhang & Lai, 2004).

Participants were requested to self-report their weight and height on the Adherence Survey in order to determine %IBW and BMI_p. The methods of determining these two values are described below.

The % IBW was determined by the following procedure:

i. Using the individual's sex, age, height, and weight, height was plotted on the appropriate CDC growth chart to determine the HFA percentile;

ii. IBW was determined according to the weight which corresponded to the same percentile as the HFA; and

iii. The %IBW was determined by dividing the actual weight by the IBW and multiplying by 100% (Zhang & Lai, 2004). See Table 3 for guidelines used to interpret HFA values.

Table 3: Interpretation of Height-For-Age Values

Status	**Height-For-Age Percentile**
Shortness/stunting	Below 3^{rd} percentile

Source: Dietitians of Canada, Canadian Paediatric Society, The College of Family Physicians of Canada, & The Community Health Nurses Association of Canada, 2004.

The BMI_p was determined by the following procedure:

i. BMI was calculated according to the following formula: weight (kg) divided by height in meters squared (m^2), multiplied by 100%;

ii. The CDC's BMI Child and Teen Calculator was used to determine BMI_p by inputting the participant's age, gender, weight (kg), and height (cm) (Zhang & Lai, 2004; Centre for Disease Control and Prevention, 2006b). See Table 4 for guidelines used to interpret BMI_p values.

Table 4: Interpretation of Body Mass Index Percentile Values

Weight Status Category	**Body Mass Index Percentile**
Underweight/Wasting	Below 5^{th} percentile
Healthy weight	5^{th} to 84^{th} percentile
At risk of overweight	85^{th} to 94^{th} percentile
Overweight	Equal to or greater than 95^{th} percentile

Sources: Centre for Disease Control and Prevention, 2006a; Dietitians of Canada, Canadian Paediatric Society, The College of Family Physicians of Canada, & The Community Health Nurses Association of Canada, 2004.

5.3.4. Eating attitudes.

The Eating Attitudes Test (EAT), developed by Garner and Garfinkle (1979), is proposed to be a self-administered tool that measures symptoms of anorexia nervosa. The original EAT, comprised of 40 questions, was developed and validated using a group of female participants with anorexia nervosa, against a group of similar controls (Pastore, Fisher, & Friedman, 1996). Since then, it has been used for screening people considered to be at a high risk for eating

disorders, but who are not diagnosed with anorexia (Garner & Garfinkel, 1979; Garner, Olmsted, Bohr, & Garfinkel, 1982). While the majority of non-clinical groups who score high do not satisfy the diagnostic criteria for anorexia nervosa, they have been identified as having disordered eating patterns which may interfere with normal psychosocial function (Garner & Garfinkle, 1979). Thus, the EAT results relate to emotional disturbances in individuals, and are independent of weight (Garner & Garfinkel, 1979; Garner et al., 1982). Participants who are anorexic tend to overestimate their body size and have EAT scores that are significantly higher than participants who over- or under-estimate their size moderately (Garner & Garfinkel, 1979; Garner et al.).

There is a shortened version of the EAT which is comprised of 26 questions, also divided into the three categories (*The Eating Attitudes Test-26 or EAT-26*) (Garner & Garfinkel, 1979; Garner et al., 1982; River Centre Clinic, n.d.). This version has been validated and it is highly predictive of the total EAT results (r=0.98) (Garner & Garfinkel, 1979; Garner et al.). Due to the proven validity of the tool, the lower subject burden, and since it has been utilized in a study of CF adults (Abbott et al., 2000), it was the tool of choice in this study.

The EAT-26 is scored by summing all the items in the survey (Garner & Garfinkel, 1979; Garner et al.). When subdivided, factor I, called dieting, is significantly related to variables concerning body image but not bulimia (Garner & Garfinkel, 1979; Garner et al.). Factor II is related to bulimia and food preoccupation (Garner & Garfinkel, 1979; Garner et al.). Factor III is negatively related to bulimia and is not significantly related to body-image concerns, being primarily related to oral control during feeding (Garner & Garfinkel, 1979; Garner et al.).

Those who score high on factor I can be classified as over-estimators who are not satisfied

with their body shape and wish to be smaller (reliability: alpha=0.90) (Garner & Garfinkel, 1979). Those who score high on factor II would be associated with a poorer outcome (Garner & Garfinkel, 1979). Those who score high on factor III tend to have a large degree of self-control when it comes to food, and also have social pressure to gain weight (Garner & Garfinkel, 1979). High scores for this factor are associated with a more positive prognosis (Garner & Garfinkel, 1979). High scores on factor III and low scores on factor II may be a better indicator of positive prognosis than the total score of all factors (Garner & Garfinkel, 1979).

White there is a child version of the EAT available, called *The Children's Eating Attitudes Test*; this tool is for use with children below the age of 15 (Smolak & Levine, 1994). Since the population was between 13 and 20 years, inclusive, using the EAT-26 was deemed appropriate for this study.

5.3.5. Body esteem.

The Body-Esteem Scale for Adolescents and Adults (BES), a 23 item, five-point Likert scale, was used to assess body esteem in the study (Mendelson, White, & Mendelson, 1997). The scale has been validated using 1334 participants, between the ages of 12 and 25, from English-speaking elementary schools, high schools, colleges, and universities in Montreal (Mendelson et al., 1997).

The scale has a total summed value as well as being broken down into three subscales. The subscales measure factors of body-esteem (BE): BE-Appearance (feelings about one's appearance), BE-Weight (feelings about one's weight), and BE-Attribution (evaluations of body and appearance from others) (Mendelson et al., 1997). Overall, the scale differentiates feelings about appearance from feelings about body weight (Mendelson et al.). Items are also worded in

such a way as to avoid having a bias towards being over or underweight. This is important with CF individuals, since they may be dissatisfied due to having a low weight rather than a high weight.

The BES is scored by adding up responses which vary from Never (0) to Always (4). Questions four, seven, nine, 11, 13, 17, 18, 19, and 21 have scores which are reversed (4 for never, 0 for always). Scores that are higher overall, and for a particular subscale as described above, are indicative of a positive body esteem (Mendelson et al., 1997).

5.3.6. Adherence.

5.3.6.1. Perceived adherence.

Participant's perceived adherence was determined via a self-designed survey entitled *The Adherence Survey* (AS). Content validity was established through a review by a dietitian and members of the thesis committee, which included a statistician. Part of this tool asked participant's to judge their adherence to physician and/or dietitian prescribed enzyme, vitamin, and nutritional supplement/s in a week. Specifically, question four assessed adherence to vitamins, question ten assessed nutritional supplements (where applicable), and questions 14 and 15 assessed adherence to enzymes for meals and snacks.

Each question was scored according to the following scheme (scores are in brackets):

a) I never take a full dose (0);

b) I take a full dose 1-2 days a week (1);

c) I take a full dose 3-4 days a week (2);

d) I take a full dose 5-6 days a week (3); and

e) I take a full dose every day of the week (4).

For this study, participant's response to the four questions in paragraph one were grouped into three categories:

1. Poor Adherence: a, b, or c was chosen;
2. Good Adherence: d was chosen; and
3. Excellent Adherence: e was chosen.

5.3.6.2. Actual adherence.

Actual adherence was determined through the Three-Day Food Diary. A column was provided in the diary for participants to record enzymes taken during meals and snacks, as well as for supplement usage. Enzymes were calculated for "Meals" if they were recorded in the Breakfast, Lunch, and Supper rows. Enzymes were calculated for "Snacks" if they were recorded in the Morning Snack, Mid-Afternoon Snack, and Evening Snack rows. Enzyme counts were totaled and the average enzyme dosage taken per single meal and snack was determined. This was also applied to supplements and vitamins for each participant, where applicable.

Recorded values were then compared to the amount prescribed from a doctor and/or dietitian, as recorded in questions 3Full, 9Full, 13M, and 13S, on the Adherence Survey. Values were interpreted as follows:

1. Adherent: If counts determined from the Three-Day Food Diary were the same as, or more than, the prescribed amounts as reported for the same treatment in the Adherence Survey.
2. Non-Adherent: If counts determined from the Three-Day Food Diary were less than the prescribed amounts as reported for the same treatment in the Adherence Survey.

In addition, adherence to dietary therapy was determined through a comparison of daily

caloric intakes from Three-Day Food Diaries to the adjusted EER (EER + 20%). Here, one was considered adherent if they consumed calories at, or above, their adjusted EER value.

5.3.7. Statistical analysis.

Microsoft Excel was used to input data, create relevant charts, and perform relevant statistical analysis.

5.4. Ethical Considerations

5.4.1. Confidentiality.

While collecting data, all tools had an identifying number (ID Code) on them which was used in order to maintain confidentiality and anonymity. The code served to keep information from each participant together. Participants were told that names should not be given on any of the survey forms and their address should not be written on the return envelope. The Communication of Results Card and the Draw Card contained participant addresses, but this information was voluntarily disclosed by participants.

As data was received, it was stored in a locked cabinet (papers) and on a password protected computer. Data will remain as such for a period of time, as specified by *The IWK Health Centre* and *The Janeway Children's Health and Rehabilitation Centre,* of five years. After that time, papers will be destroyed and computer files will be deleted. All envelopes were destroyed upon receipt. Only the researcher and the researcher's supervisor had access to the data.

If data is published, there will be no way to identify participants when results are written up and published. No names or ID Codes will be referred to.

5.4.2. Ethics approval.

Ethics approval was obtained from *The Mount Saint Vincent University Ethics Review Board,*

The IWK Research Ethics Board, and *The Memorial University Human Investigation Committee* (for *The Janeway Children's Health and Rehabilitation Centre*), including amendments that were made. As well, *Debriefing Forms* (Appendix D and E) were used for participants from both hospitals, as specified by the institutions.

5.5. Advantages and Limitations of Methodology

Surveys/questionnaires have the advantage of being easy to administer, with little respondent burden (Mendelson et al., 2001). However, problems may occur with the self-disclosure of accurate information as participants may be hesitant to admit that they do not adhere to recommendations. The use of physician reports has been found to be no more accurate in predicting adherence than chance, as participants may be more resistant to admit non-adherence to their doctor than a researcher (Lask, 1994). By being cognizant of question wording, this potential problem was minimized. It was stated on the AS that non-adherence is common in the CF population, and the researcher is wondering if, and why, they are non-adherent. Focus groups and interviews are not feasible for this population, due to the risk of transmission of bacteria from person to person (CCFF, 2003).

Measurements of dietary intake rely on memory, which is not perfect (Livingstone & Robson, 2000). Other errors, which may be related to memory or intentional manipulation include under-reporting, omitting foods, over reporting, and reporting foods that were not actually eaten (phantom foods) (Livingstone & Robson, 2000). Some other factors which may effect recall include an excess of information given to the participant, providing visual pictures of foods, training participants in completing the questionnaires, the salience of food items (it is often easier to remember main course items), and food frequency, common foods are more likely

to be recalled (Baranowski et al., 1986; Emmons & Hayes, as cited in Livingstone & Robson, 2000).

Dietary studies conducted on children and adolescents, when determining energy intake, report a between-subject variation on recall of 20% or more (Livingstone & Robson, 2000). Fortunately, there are a few things that can be taken into consideration when determining dietary intake in adolescents. Methods with a low respondent burden are more likely to be adhered to, such as the 24 hour recall or a three-day food diary/record, when compared to weighted records or those for longer periods of time (Livingstone & Robson, 2000). Recalls are often associated with problems estimating portion sizes, which can be lessened by giving handouts with sample portion size equivalents (Livingstone & Robson, 2000). Lastly, teenagers have distinct eating patterns versus adults, such as snacking often, grazing, or skipping meals (Livingstone & Robson, 2000). These characteristics were taken into consideration when developing the record form, by including spaces for snacks and not just meals.

Section 6: Results

6.1. Response Rate

Adolescents and young adults aged 13 to 20 years, inclusive, who attended CF clinics at the IWK and the Janeway made up the sampling frame for the study. According to the 2002 Canadian CF Patient Data Registry, there are 73 patients between the ages of 12 and 17, and 65 between the ages of 18 and 23 located in eastern Canada (CCFF, 2002). Therefore, a conservative estimate of the total population of Canadians, between the ages of 13 to 20, with CF living in the Atlantic Provinces is 80. Initially, participants were recruited who fell into the age range of 13 to 17, inclusive. Twenty-three packages were sent to possible participants from the IWK, and ten to the Janeway. These 33 possible participants received a follow-up letter approximately one month after packages were mailed. After receiving few returns, an amendment was sent to the appropriate ethics boards in order to change the age of inclusion to 13 to 20, inclusive. Once approved, packages were sent to 12 additional potential participants from the IWK, and six from the Janeway.

In total, there were 16 potential participants from the Janeway (31% of total sample size) and 35 (69%) from the IWK, giving a total sample size of 51 possible participants. A total of nine responses were collected, giving a return rate of 17.6%. The IWK accounted for 78% of the returns, versus 22% from the Janeway. The low response rate is attributed to the fact that this population is young and may not have been highly motivated to complete the questionnaires. While of modest length, time may also have been a factor in participation.

6.2. Description of the Respondents

The demographic profile (Questions 19 and 20 on the Adherence Survey) was designed to obtain information about gender and age. Clinic location of the respondents was determined via the ID code on each participant's forms. Self-reported anthropometric data (Questions 21 and 22 on the Adherence Survey) was collected to provide information for calculating the nutritional status of participants. The gender profile of respondents was evenly matched, at four male participants and five female. It was possible for participants from the IWK to reside in the province of Nova Scotia, New Brunswick, or Prince Edward Island. Those from the Janeway would typically reside in Newfoundland and Labrador. Taking this into consideration, along with the fact that 69% of potential participants were from the IWK, it is not extraordinary that the majority of respondents attended the IWK clinic (78%). All respondents were included in the calculations, except where specified. See Table 5 for a summary of the study participants.

Table 5: Summary of Characteristics of Study Participants

Total Possible Participants	51
Total Participants	9
Return Rate	17.6%
Number of Male Participants	4
Number of Female Participants	5
Number of Participants from the Janeway	2
Number of Participants from the IWK	7
Mean Age	15.2 years

6.3. Nutritional Intake

6.3.1. Coding and interpretation of information.

Nutritional intake was recorded by all nine participants via the Three-Day Food Diary. Despite the inclusion of detailed instructions for the completion of food diaries, many lacked adequate descriptions. To compensate for this, the following strategies were utilized for each participant record:

a) If item amounts were given for a food at one time, but not subsequent times within the record, the amount used initially was recorded thereafter.

b) In instances where the fat percentage of milk was not given throughout the entire record, it was assumed that whole milk was used. This is due to the fact that those with CF are counseled to consume a diet rich in fat and energy.

c) In instances where yogurt was consumed but type not given, yogurt made with whole milk was recorded due to the reason given in b.

d) In instances where amounts of a food were not given throughout the entire record, typical serving sizes according to Canada's Food Guide to Healthy Eating and the Canadian Diabetes Association's Handy Portion Guide were used for all appearances of the item. See Table 6 for a listing of serving sizes (Canadian Diabetes Association, 2005; Health Canada, 2005a).

e) In instances where meat cuts were not provided, conservative estimates of cuts were used. These were used consistently across all responses that lacked adequate descriptions. Table 7 provides a breakdown of used cuts. In all cases, fattier meats were chosen from possible choices in the database, due to the reason given in strategy b.

Table 6: Interpretation of Quantities of Food Items Listed in Participant Three-Day Food Diaries

Food Group Items	**Amount Used**
Serving of meat/poultry	3 oz
One (1) bowl or plate of pasta or rice	½ cup
One (1) glass of fluid/beverage	1 cup/8 oz.
One (1) piece fruit/vegetable	1 medium sized piece
One (1) handful	1 cup
One (1) bowl of yogurt	¾ cup
One (1) bowl of hot cereal	¾ cup
One (1) bowl of cold cereal	30 g
Peanut butter	2 tbsp.
One (1) bowl of salad or vegetables	1 cup

Sources: Health Canada 2005b; Canadian Diabetes Association, 2005.

Table 7: Interpretation of Meat Product Cuts Listed in Participant Three-Day Food Diaries

Type of Meat Listed [a]	**Selection from USDA Database Recorded** [b]
Chicken: • Cut= Thigh, wing, leg, drumstick, breast	• Chicken, broilers or fryers, (cut), meat and skin, cooked, roasted
Turkey	• Turkey, all classes, light meat, cooked, roasted
Pork: • Chop • Roast	• Pork, fresh, loin, center rib (chops), boneless, separable lean and fat, cooked, braised • Pork, fresh, loin, center rib (roast), boneless, separable lean and fat, cooked, roasted
Beef: • Ground • Steak • Roast	• Beef, ground, 70% lean/30% fat, crumbles, cooked, pan-browned • Beef, top sirloin, separable lean and fat, trimmed to 1/8" fat, all grades, cooked, broiled • Beef, round, bottom round, separable lean and fat, trimmed to 1/8" fat, all grades, cooked, roasted.

a. Meat listed in participant Three-Day Food Diaries without specific details as to cut.
b. USDA Nutrient Database for Windows- HealtheTech Search SR-18 program used for nutrition analysis (USDA Nutrient Data Products and Services, 2006).

6.3.2. Nutrient intakes.

Average daily calorie, fat, carbohydrate, and protein intakes were determined for all nine participants, according to nutrient analysis from completed Three-Day Food Diaries. Daily caloric intakes ranged from approximately 1042 to 2748 Cal a day, with a mean of 2174 Cal; daily fat intake ranged from approximately 43 to 139 g a day, with a mean of 83 g; daily carbohydrate intake ranged from approximately 102 to 354 g a day, with a mean of 244 g; and daily protein intake ranged from approximately 54 to 102 g a day, with a mean of 82 g. Calculating the caloric contribution of each major nutrient determined that the mean percent of

calories contributed from fat was 34.6%, that from carbohydrates was 48.65%, and that from protein was 16.08%. See Appendix N for a detailed list of each participant's nutritional intake.

6.3.3. Estimated Energy Requirements.

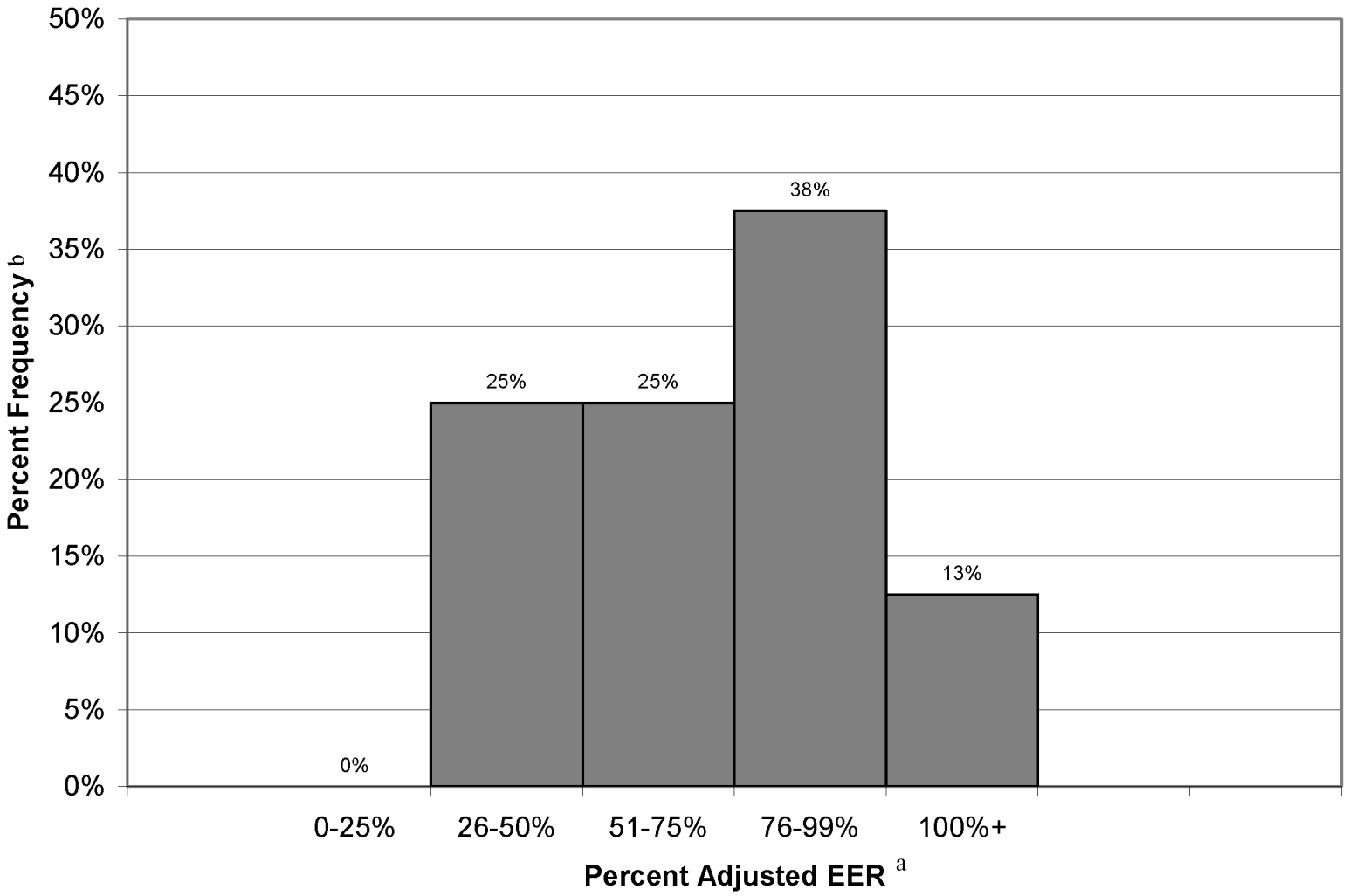
Estimated Energy Requirements were calculated using formulas given in Section 5.3.2. Since daily physical activity was not assessed in this study, a conservative estimate for the physical activity coefficient (PAC) was used in all cases. The presence of CF may make engaging in strenuous or prolonged periods of physical activity difficult. Therefore, it is unlikely that participants fell into the Very Active category for the majority of time. However, given the fact that all participants were relatively young, Low Active was used as the estimate for the PAC. Therefore, a conservative EER was calculated for eight participants, with one participant failing to provide adequate data (height and weight omitted). Given the assumption that participants fell into the Low Active category, this value represents the lowest daily caloric intake which should be consumed by the participants, not taking into consideration the effects of the disease. Having an intake less than this value may be problematic for the participants.

Given the fact that this population has CF, it is recommended that 20-50% more calories should be consumed per day than the EER (see Section 3.5.2. for more information). Adjusted EER's were calculated for the eight participants, adding an additional 20% onto the previously calculated EER. This value, under the assumption that participants fell into the Low Active category, represents the lowest preferred or recommended daily caloric intake. Those achieving an intake of the adjusted EER or more would be considered to have an adequate caloric intake. After calculating each participant's EER and adjusted EER, the frequency distributions were determined and are graphically depicted in Figures 7 and 8.

As seen in Figure 7, half of the participants met or exceeded their EER's. This indicates that half of the participants (n=4) did not meet the lowest daily caloric intake recommended, not taking into consideration their health status.

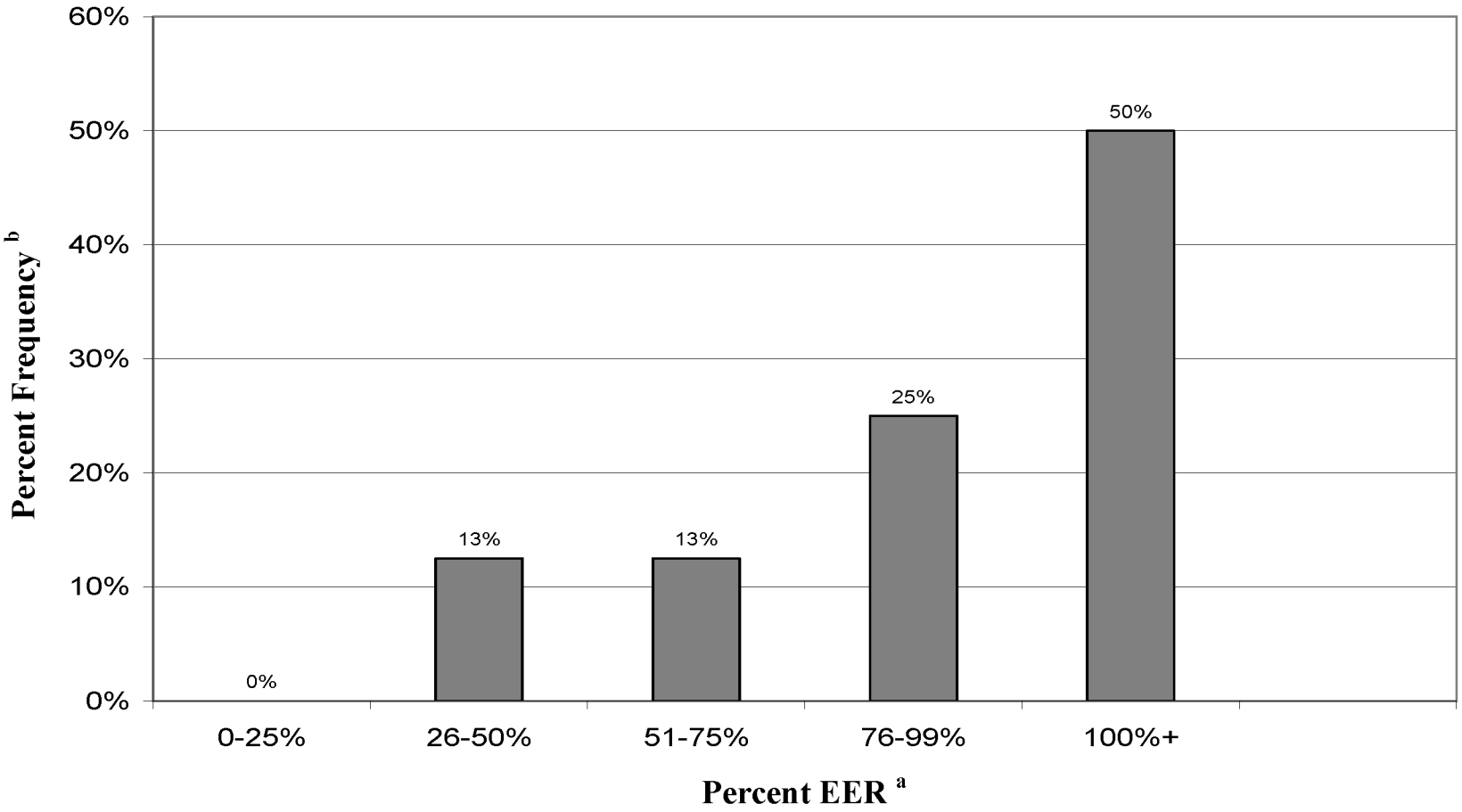
As can be seen from Figure 6, half of the participants (n=4) had an average daily caloric intake which was 76% of their adjusted EER or more. However, only 13% had an intake which was 100% or more. While no participants had a daily caloric intake of 25% or less of their adjusted EER, the majority are not meeting the lowest preferred, or recommended, daily caloric intake of 120% EER.

Figure 6: Distribution of Caloric Intakes when Compared to Adjusted Estimated Energy Requirements (EER)



a. Adjusted EER= EER + 20%. b. n=8.

Figure 7: Distribution of Caloric Intakes when Compared to Estimated Energy Requirements (EER)



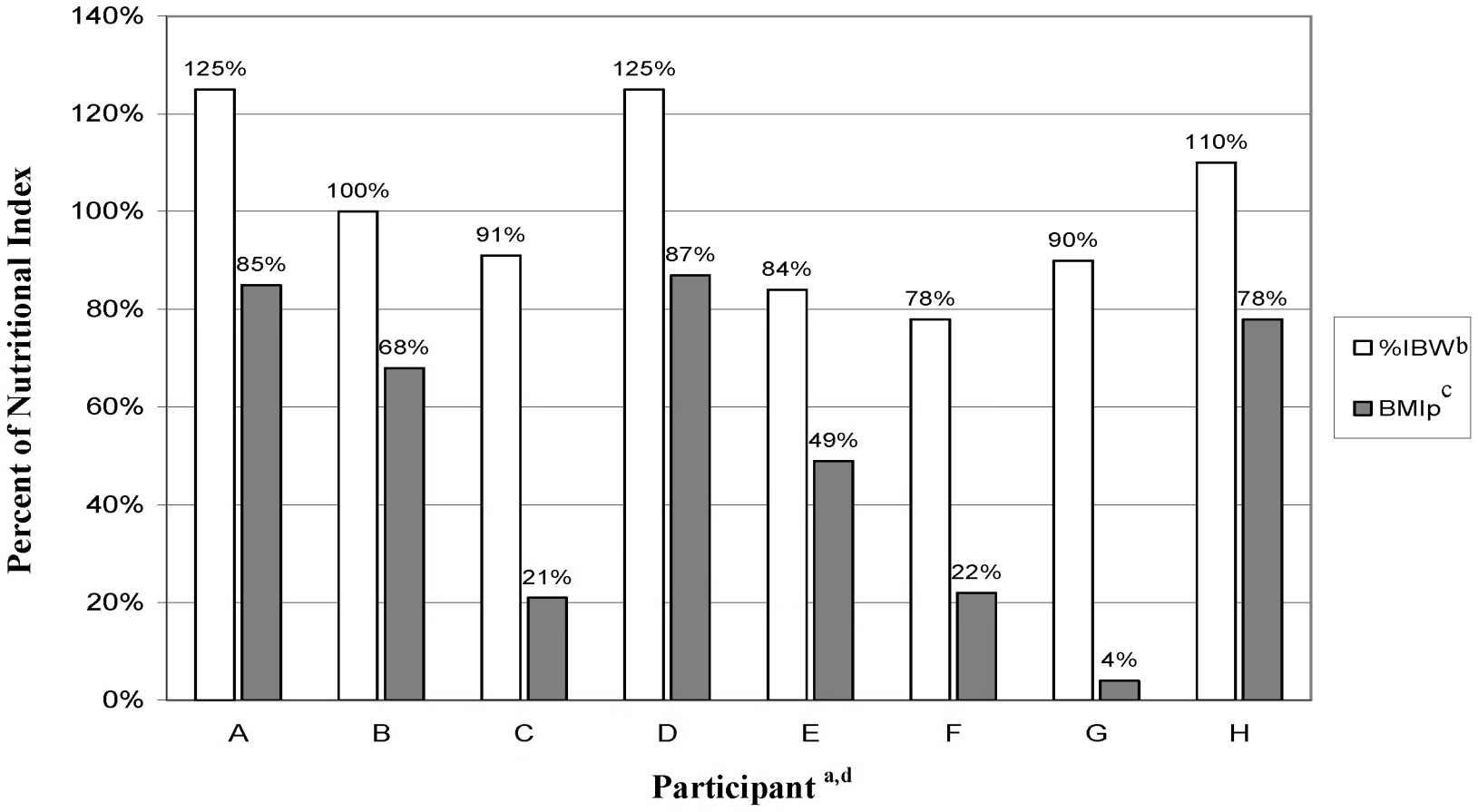
a. Formula for EER: Health Canada, 2005b.
b. n=8.

6.3.4. Nutritional status.

The nutritional status of participants was determined by calculating %IBW and BMI_p, as described in Section 5.3.3. Both calculations were determined for eight participants, since one did not self-report weight and height. Percent IBW, rounded to the nearest number, ranged from a low of 78% to a high of 125%, with a mean of 100%. Half of the participants fell into the 75 to 99% IBW range, inclusive, and the other half fell into the 100% or above range.

When looking at the BMI_p, participants ranged from a low of the fourth percentile to a high of the 87^{th}, with a mean at the 52^{nd} percentile (values rounded to the nearest number). According to the BMI_p, 25% of participants were classified as being at risk of overweight, 63% were in the healthy status category, and 13% were underweight. It should be noted, however, that since the population is rather small, the 13% who were underweight is equivalent to one participant, who also falls into the underweight or wasting category. At the same time, those who were classified as at risk for becoming overweight is equivalent to two participants. See Figure 8 for an overview of participant %IBW and BMI_p values.

Figure 8: Nutritional Status of Participants



a. In order to maintain participant confidentiality, the identifying labels used in this graph were assigned randomly and bear no relation to identifiers used in previous tables and charts

b. %IBW= Percent Ideal Body Weight.

c. BMI_p= Body Mass Index Percent. d. n=8.

6.4. Assessing Body Esteem

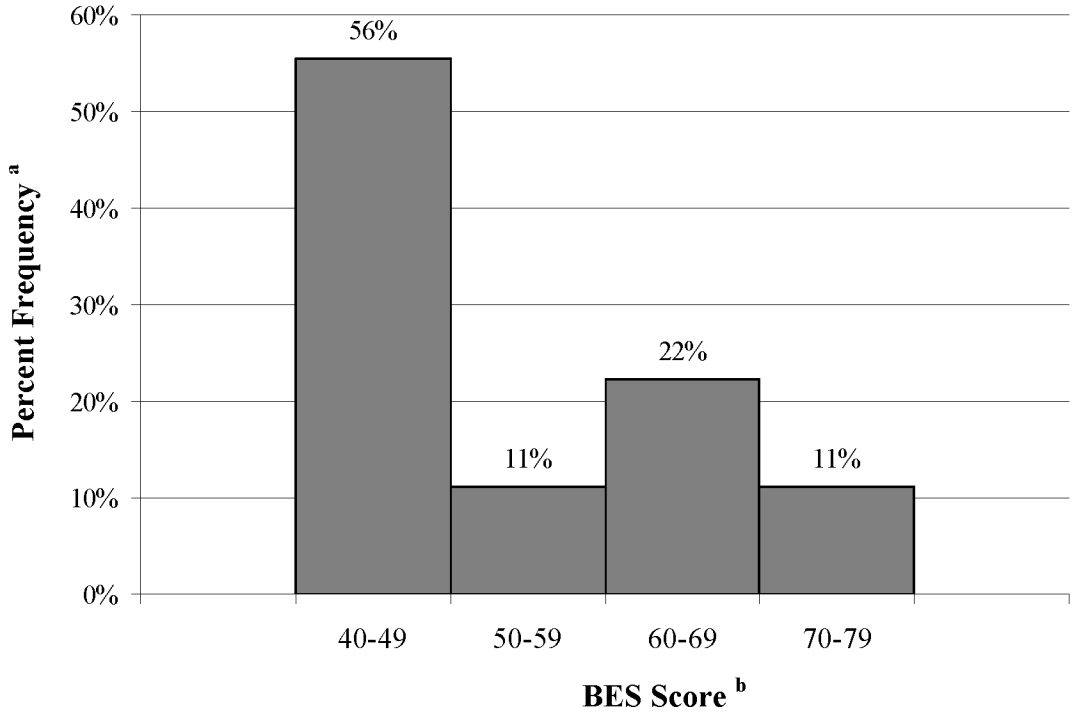
Body esteem was assessed using the *Body Esteem Scale for Adolescents and Adults* (BES). As described in Section 5.3.5., the higher the BES score, the more positive one's body esteem. Therefore, following the scoring scheme for the scale, those who are in category four (between 70-79) have the highest BES score out of all participants, and thus the most positive body esteem.

While the scoring mechanism does not have a particular cut-off point, the lowest possible score one could obtain on the scale is zero, and the highest is 92. For this study, BES scores were calculated for all nine participants and broken up into four categories as follows:

1. Scores between 40-49;
2. Scores between 50-59;
3. Scores between 60-69; and
4. Scores between 70-79.

Scores ranged from 41 to 74, with a mean of 52.67. The majority of participants (56%) had scores in the range of 40 to 49, inclusive, with 11% obtaining scores in the range of 70 to 79, inclusive. When compared to the highest obtainable score of 92, the majority of participants fell in the 43.48 to 53.26% range. The highest individual score of 74 is 80.43% of the highest possible score. By comparison, the lowest individual score of 41 is 44.57% of the highest possible score. Therefore, there were no participants who scored much below half of the highest possible score. See Figure 9 for a summary of overall participant BES scores.

Figure 9: Distribution of Body Esteem Scale (BES) Scores.



a. n=9.
b. Higher scores indicative of more positive body esteem (Mendelson et al., 2001).

When the BES was divided into its three subscales, the mean score for BE appearance was 57.23% of the highest possible score for that subscale, the mean score for BE weight was 54.16% of the highest possible score, and the mean score for BE attribution was 62.2% of the highest possible score. This indicates that in general, the participants were at the half way mark or above with respect to each body esteem related subscale. For more detailed information about the three subscales, see Table 8 below.

Table 8: Body Esteem (BE) Scale Subscale Scores

Subscale	Lowest Possible Score	Highest Possible Score[b]	Range of Scores	Mean Score[a]
BE Appearance	0	40	16-34	22.89
BE Weight	0	32	5-31	17.33
BE Attribution	0	20	10-15	12.44

a. n=9.
b. High scores indicative of more positive body esteem (Mendelson et al., 2001).

6.5. Assessing Eating Attitudes

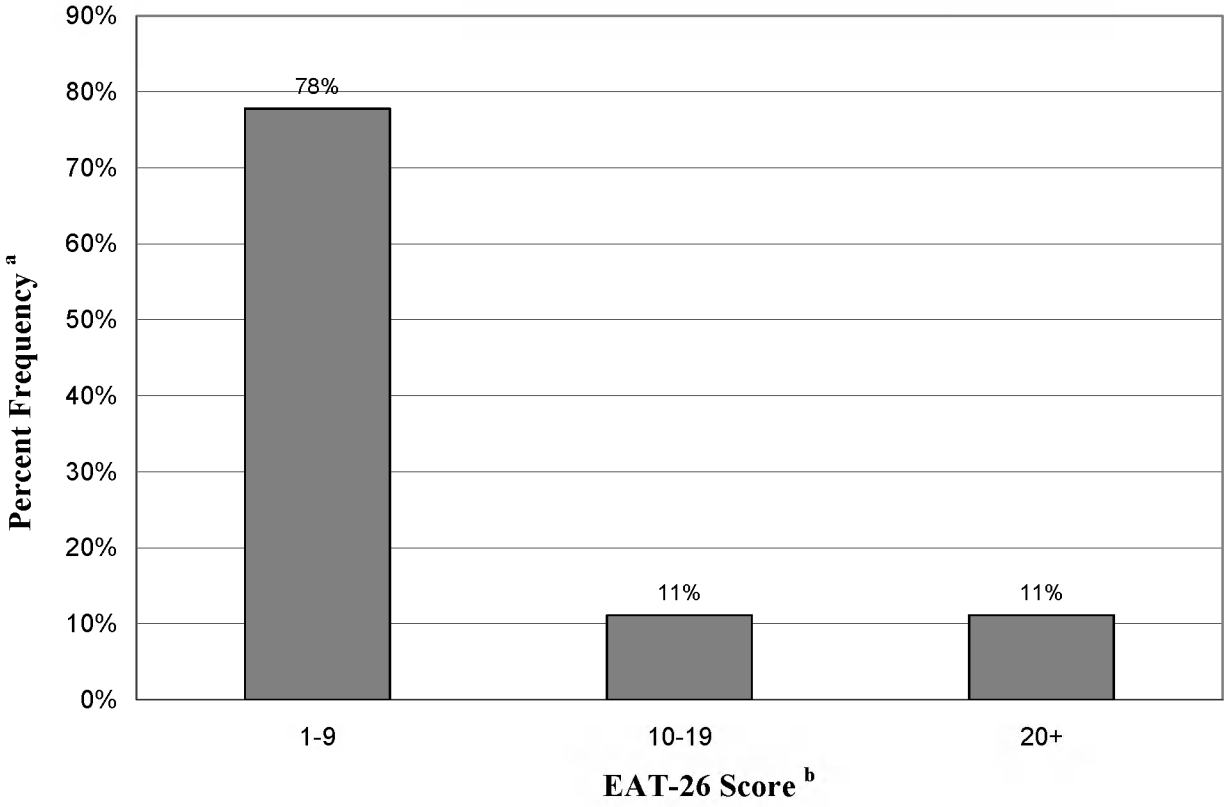
Eating attitudes were assessed via EAT-26, as described in Section 5.3.4. Scores were determined for all nine participants, with a range of responses from three to 21, and a mean of 7.78. The lowest possible score for the EAT-26 is zero and the highest is 78. Unlike the BES, the lower the score on the EAT-26, the more positive one's eating attitudes. As stated in category three of Section 5.3.4., values at or above 20 indicate the individual is at risk for developing an eating disorder (Garner & Garfinkle, 1979).

For this study, participant EAT-26 values were totaled and broken up into three categories as follows:

1. Values between 1-9: considered low risk for comparison purposes by investigator, but not identified as such by Garner & Garfinkle (1979).
2. Values between 10-19: considered medium risk for comparison purposes by investigator, but not identified as such by Garner & Garfinkle (1979).
3. Values at 20 or above: considered to be at risk (high risk) for disordered eating behaviors, according to the scoring of the tool (Eating Attitudes Test-Eating Disorder, 1999).

The majority of participants, with scores in the range of one to nine (78%), were not considered at risk for the development of disordered eating behaviors. One participant did fall into the at risk category, having a score above 20. See Figure 10 for a summary of EAT-26 scores.

Figure 10: Distribution of Eating Attitudes Test-26 (EAT-26) Scores



a. n=9.
b. Scores at or above 20 at risk for developing disordered eating attitudes (Garner & Garfinkle, 1979).

Similarly to the BES, the EAT-26 can be divided into three subscales: dieting, bulimia and food preoccupation, and oral control. Most participants ranged fairly low in each subscale, indicating a positive eating attitude with the absence of dieting behaviors. However, scores were

higher for the oral control subscale, with one participant scoring 61.90% of the highest possible score. On average, the scores were still below half of the highest possible score. See Table 9 for a summary of subscale scores.

Table 9: Eating Attitudes Test-26 Subscale Scores

Subscale	**Lowest Possible Score**	**Highest Possible Score**	**Range of Scores**	**Mean Score[a,b]**
Dieting	0	39	0-5	1.67
Bulimia and Food Preoccupation	0	18	0-7	1.67
Oral Control	0	21	2-13	4.44

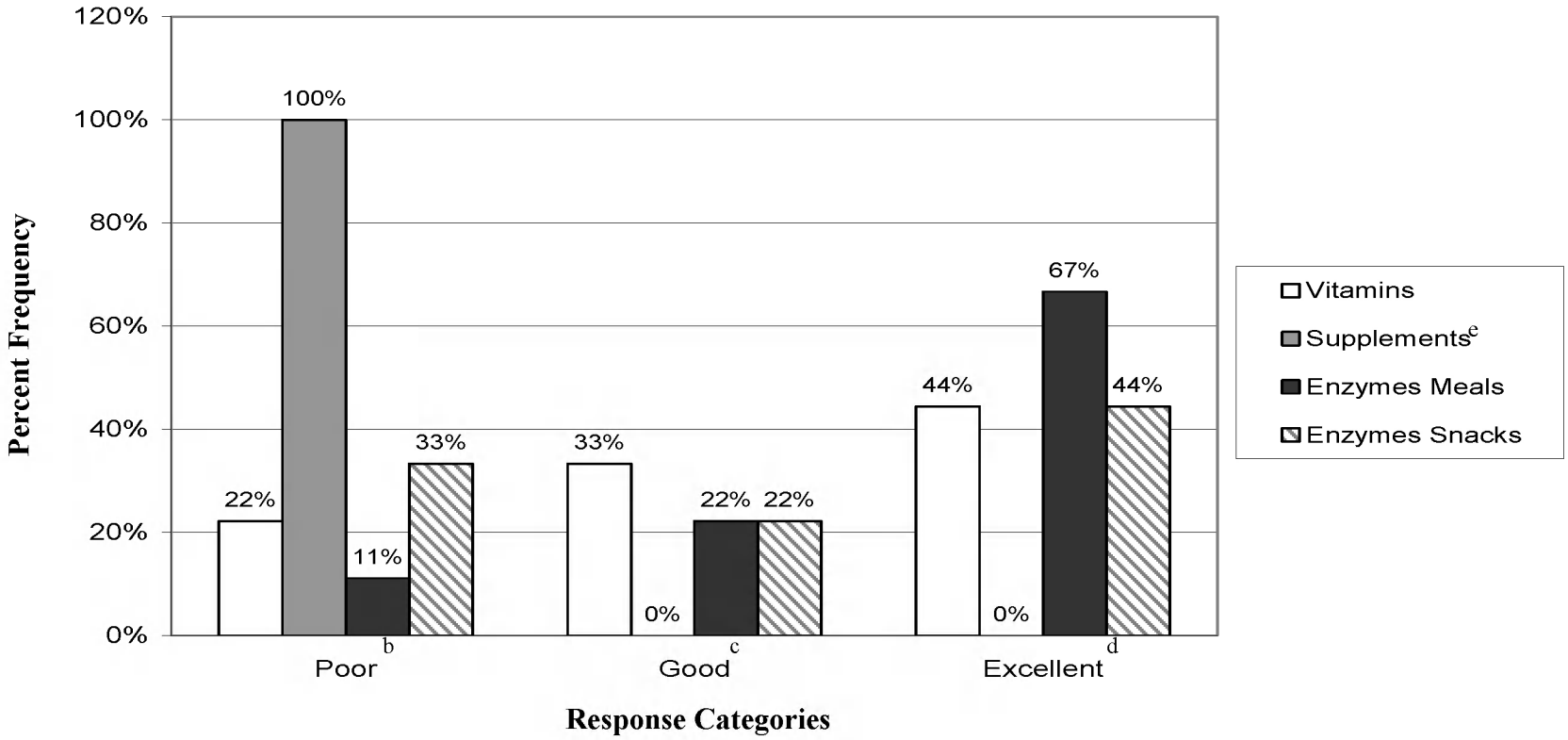
a. n=9.
b. Scores at or above 20 at risk for developing eating disorder (Garner & Garfinkle, 1979).

6.6. Assessing Adherence

Data was obtained on the Adherence Survey from nine participants, all of whom (on Questions 1 and 13) had been prescribed enzymes and vitamins, and four of whom had been prescribed nutritional supplements (Question 7).

As described in Section 5.3.6.1., perceived adherence to treatment was determined via the AS. The majority of participants rated themselves as being in the Excellent category with respect to adherence to vitamins (44%) and enzymes taken with meals (67%). The four participants who were prescribed nutritional supplements rated themselves as being in the Poor category. Despite a high perceived adherence level for enzymes taken with meals, 20% fewer participants rated themselves as being in the Excellent category with respect to adherence to enzymes with snacks. For a graphical representation of perceived adherence to treatment, as determined by the AS, see Figure 11.

Figure 11: Distribution of Perceived Treatment Adherence Scores[a]



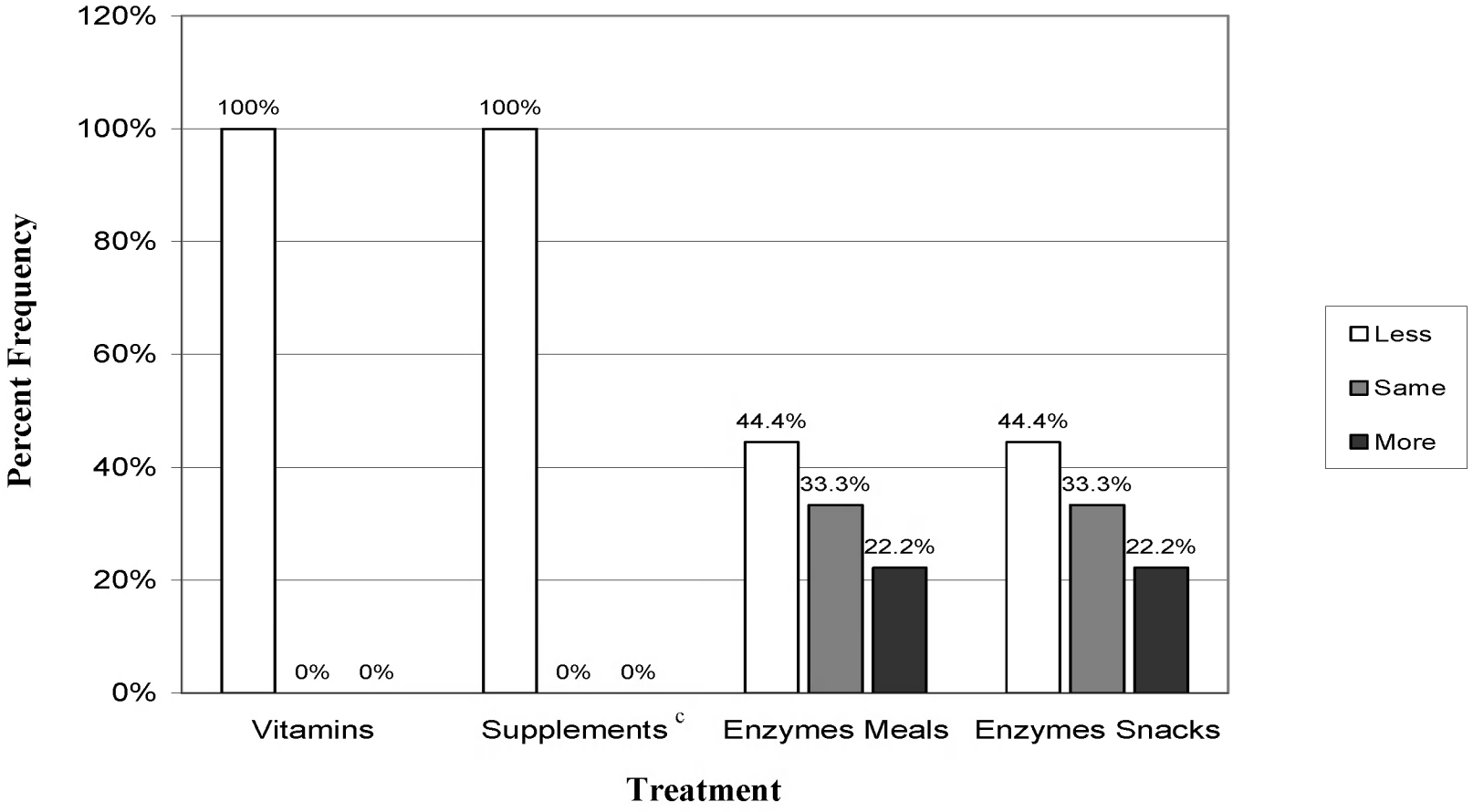
a. Perceived adherence assessed via Adherence Survey.
b. Participant reported they take a full dose of treatment four days a week or less.
c. Participant reported they take a full dose of treatment five or six days a week.
d. Participant reported they take a full dose of treatment every day of the week.
e. n=4 for supplements, n=9 for all other treatments.

Actual treatment adherence, as described in Section 5.3.6.2., was assessed through a comparison of values recorded on Three-Day Food Diaries and amounts prescribed from a doctor/dietitian, as recorded on the AS. Nine participant records were included for all calculations except for nutritional supplements (n=4). Results for actual treatment adherence with enzymes taken with meals and snacks were identical, with 44% taking less than prescribed, 33% taking the same amount as prescribed, and 22% taking more. Out of all the participants who were prescribed vitamins (n=9), all took less than prescribed each day. Similarly, all four participants who were prescribed nutritional supplements took less than prescribed. See Figure 12 for more information.

In determining an overall level of treatment adherence for participants, those who took the same amount or more of a prescribed treatment were considered adherent. Since actual treatment adherence was poor for vitamins and supplements, Figure 13 depicts actual adherence levels for enzymes, in terms of the two adherence categories.

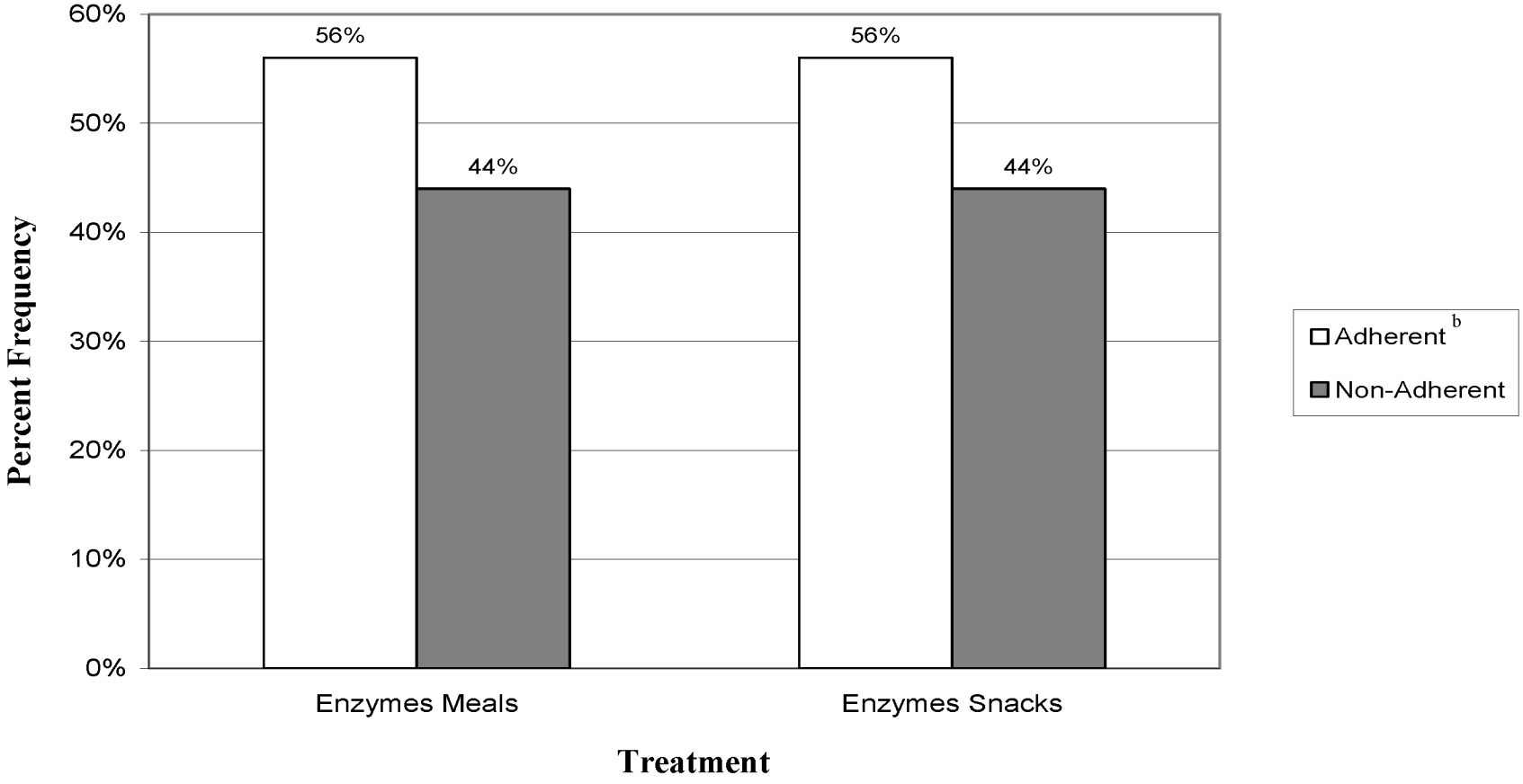
Nutrient intakes and proportions of daily adjusted EER, previously described in section 6.3.2 and 6.3.3, were used to categorize participants into "adherent" and "non-adherent" categories. As can be seen in Figure 6, the majority of participants did not meet the recommended adjusted EER. Therefore, those that did not meet this value were considered non-adherent, and those who did were considered adherent. A total of one participant (13%) was considered adherent to caloric recommendations. See Figure 14 for more information.

Figure 12: Comparison of Amounts Consumed[a] of a Treatment versus Amounts Prescribed[b]



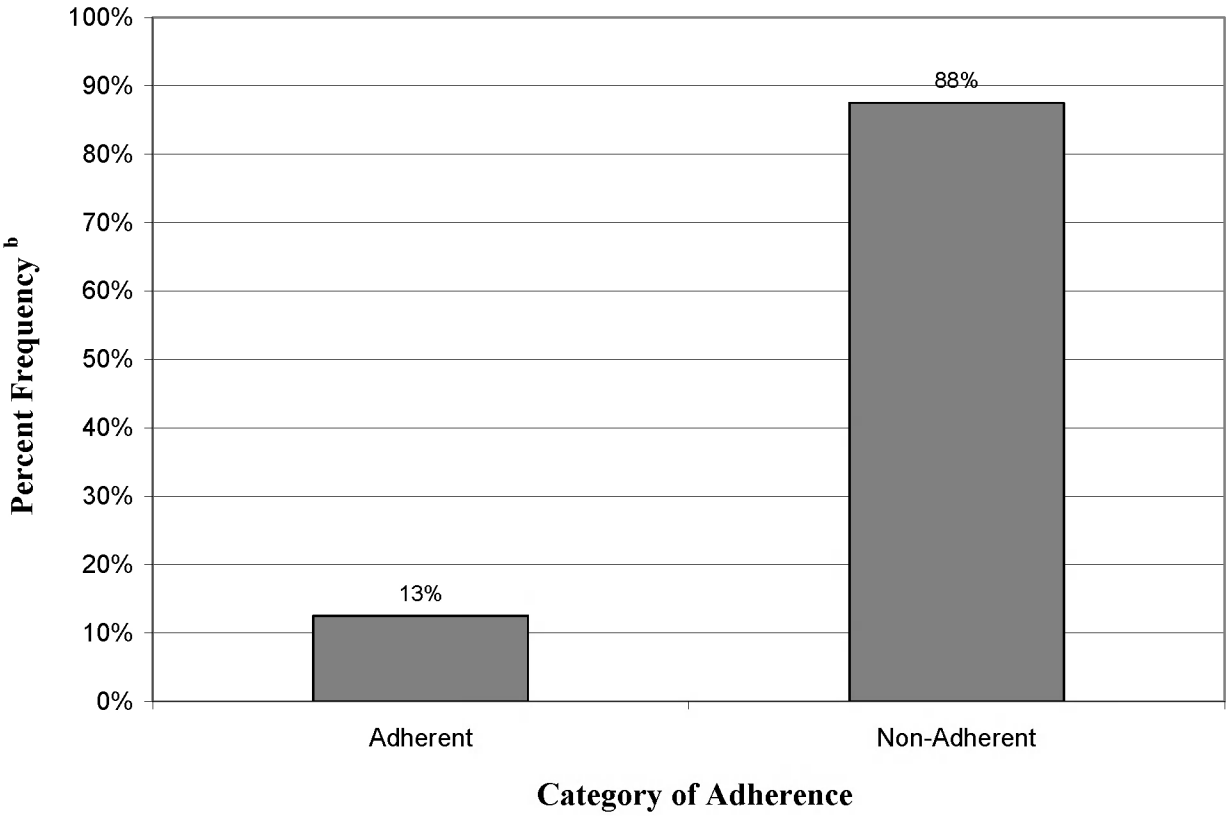
a. As determined via Three-Day Food Diary.
b. As determined by questions 3Full, 9Full, 13M, and 13S on Adherence Survey.
c. n=4 for supplements, n=9 for all others.

Figure 13: Distribution of Actual Enzyme Adherence Levels[a]



a. n=9.
b. Participants considered adherent is so for enzymes taken with meals and snacks.

Figure 14: Distribution of Adherence to Dietary Recommendations[a]



a. Optimal dietary recommendations used were equivalent to the adjusted EER (EER + 20%).
b. n=8.

For comparison purposes, the following are a set of tables comparing perceived adherence as taken from the Adherence Survey to actual adherence, as determined from criteria listed above.

Table 10: Comparison of Perceived Versus Actual Adherence for Vitamins

Participant[a]	Perceived Adherence[b]	Actual Adherence[c]
A	Good	Non-Adherent
B	Excellent	Non-Adherent
C	Poor	Non-Adherent
D	Good	Non-Adherent
E	Good	Non-Adherent
F	Excellent	Non-Adherent
G	Poor	Non-Adherent
H	Excellent	Non-Adherent
I	Excellent	Non-Adherent

a. In order to maintain participant confidentiality, the identifying labels used in this graph were assigned randomly and bear no relation to identifiers used in previous tables and charts.
b. Determined via Adherence Survey.
c. Determined from comparison of amount consumed from Three-Day Food Diary and amount prescribed from Adherence Survey (Questions 3Full, 9Full, 13M, and 13S).

Table 11: Comparison of Perceived Versus Actual Adherence for Enzymes Taken with Meals

Participant[a]	Perceived Adherence[b]	Actual Adherence[c]
A	Good	Non-Adherent
B	Excellent	Adherent
C	Excellent	Adherent
D	Poor	Non-Adherent
E	Excellent	Non-Adherent
F	Excellent	Adherent
G	Good	Adherent
H	Excellent	Non-Adherent
I	Excellent	Adherent

a. In order to maintain participant confidentiality, the identifying labels used in this graph were assigned randomly and bear no relation to identifiers used in previous tables and charts.
b. Determined via Adherence Survey.
c. Determined from comparison of amount consumed from Three-Day Food Diary and amount prescribed from Adherence Survey (Questions 3Full, 9Full, 13M, and 13S).

6.7. Relationships between Assessed Variables

To try to determine if and how adherence is related to body esteem, eating attitudes, and to the measures used to determine nutritional adequacy, participants were grouped into two

categories, again labeled "Adherent" and "Non-adherent". Since no participant was adherent for vitamins or supplements, only the enzyme levels for meals and snacks were used. A participant was considered adherent if they were adherent to physician-prescribed enzyme dosages taken with meals *and* snacks.

Two-tailed independent sample t-tests were calculated to determine if there were any significant differences between the mean scores of the two adherence groups for each of the listed response variables. Results, as shown in Table 10, indicate no significant differences were found. Therefore; in the population of all adolescents with CF, based on the results of this study, one cannot predict differences in BES, EAT-26, EER, BMI_p or %IBW based on the individual's adherence category.

Despite the fact that results are not statistically significant, looking at the values show that those who were adherent had a more positive body esteem, had a more positive eating attitude, and had a higher BMI_p, and %IBW than those who were non-adherent. The participants who were adherent had a higher body weight than those who were non-adherent, despite the fact that they seemed to consume less calories.

Table 12: Relationships between Body Esteem, Eating Attitudes, Nutritional Status, and Nutritional Intake by Adherence Level[a]

Enzyme Adherence Category	Count	Body Esteem Scale Score (M ± SD)[b,d]	Eating Attitudes Test-26 Score (M ± SD)[e]	Percent Adjusted Estimated Energy Requirement (M ± SD)	Body Mass Index Percentile (M ± SD)	Percent Ideal Body Weight (M ± SD)
Adherent[c]	4	57.75 ± 14.27	5.75 ± 2.5	60.94% ± 23.90%	63.5% ± 29.38%	106.52% ± 14.62%
Non-Adherent	5	48.6 ± 10.31	9.4 ± 7.37	75.92% ± 34.24%	40% ± 35.24%	94.05% ± 20.91%

a. Determined via two-tailed independent sample t-tests.
b. Mean ± Standard Deviation.
c. Participants considered adherent is so for enzymes taken with meals and snacks.
d. High scores indicative of more positive body esteem (Mendelson et al., 2001).
e. Scores at or above 20 at risk for developing eating disorder (Garner & Garfinkle, 1979).

Section 7: Discussion

This study was designed to investigate the relationship between adherence and body esteem in adolescents and young adults with cystic fibrosis. The discussion that follows will focus on the following aspects raised by the findings of the study: nutritional intake, nutritional status, body esteem, eating attitudes, adherence, and the application of the theoretical framework to the study.

7.1. Nutritional Intake

The caloric requirements for those with CF are approximately 120 to 150% of recommendations for similar healthy peers (Roy, Darling, & Weber, as cited in Pencharz & Durie, 1993; Pencharz et al., as cited in UK Cystic Fibrosis Trust Nutrition Working Group, 2002; Sinaasappel et al., 2002; Creveling et al., 1997). Reasons for this increased requirement, as explained in Sections 3.2.3 to 3.2.5., include: an increased loss of energy due to malabsorption and maldigestion (Durie et al., 1998), an increased energy expenditure due to the work of breathing and coughing (Pencharz, Hill, Archibald, Levi, & Newth, as cited in Pencharz & Durie, 1993; Sinaasappel et al., 2002; Marin et al., 2004), and the presence of secondary complications such as glucose intolerance, diabetes mellitus (Creveling et al.; Littlewood, 2002), and an impaired liver function (Sinaasppel et al., 2002).

Despite the extra energy demands on the CF body, poor intake is common in this population (Collins et al., 1998). Due to the imbalance between energy needs and dietary intake, studies have reported a prevalence of malnutrition of between 34 and 59% (Lai et al., 1998; Ionescu et al., 1998, 2000; Dray et al., 2004; Steinkamp & Wiedemann, 2002). While many aspects of CF are beyond the individual's control, nutrition is one factor over which power can be exercised.

The failure to maintain an optimal nutritional status/intake can result in problems such as: impaired growth (Mearns, as cited in Sinaasappel et al., 2002), a decline in lung function (Steinkamp & Wiedmann, 2002; Konstan et al., 2003), osteoporosis/osteopenia (Conway et al., 2000; Fok et al., 2002), and delayed puberty (UK Cystic Fibrosis Trust Nutrition Working Group, 2002; Pencharz & Durie, 1993). For more information, see Section 3.4.

When interpreting and drawing conclusions based on the Three-Day Food Diaries, it is important to note that many did not contain adequate data. Section 6.2.1. details the strategies used to record nutrient intakes and analyze the information provided. Despite these strategies, it is possible that certain items were not estimated correctly. As a result, true daily caloric intakes of participants may vary from those determined in this study. Measurements of dietary intake are subject to errors caused by a faulty memory, or the intentional under-reporting and/or over-reporting of food items (Livingstone & Robson, 2000). Dietary studies conducted on children and adolescents report a between-subject variation on recall of 20% or more (Livingstone & Robson, 2000).

Given these limitations, results from this study indicate that the participants had a mean daily caloric intake of 2174 Cal. This is comparable to results from various studies in which participants averaged between 2200 to 2345 Cal a day (Ionescu et al., 2002; Steinkamp & Wiedmann, 2002; Marin et al., 2004; Walkowiak & Przyslawski, 2003; Stark et al., 1997; Collins et al., 1998). When caloric intakes were contrasted to EER's, half (total sample of 8 participants) met or went beyond the value. However, when the adjusted EER were determined, only one participant (equivalent to 13% of the population) met or exceeded the value, with the majority (50%) consuming 76 to 99% of their adjusted EER. In theory, the adjusted EER

represents a modest clinical recommendation for caloric intake in this population.

It is recommended that the contribution of daily calories from fat should be between 30-40% (Pencharz et al., as cited in Pencharz & Durie, 1993; Marin et al., 2004). The mean percentage of calories contributed by fat in this study was 34.6%, which falls into the recommended range. The majority of participants (78%) met the recommended fat intake.

The relationship between dietary intake (percent of adjusted EER) and adherence was determined via two-tailed independent sample t-tests (see Table 13). For this statistical comparison, adherence was defined as those participants who were adherent to enzymes for both meals and snacks (four adherent, five non-adherent). Results indicate that there was no statistical significance between adherence level and caloric intake. However, those who were adherent tended to have a lower caloric intake then those who were non-adherent. While those who are adherent may in fact have consumed less of their recommended calories, the following factors, if present, may have affected results: information on Three-Day Food Diaries of those who were considered adherent was incomplete, over-reporting of information on Three-Day Food Diaries of those who were non-adherent, misinterpretation of incomplete data as discussed in Section 6.3.1., and the participant's weight and/or height may have been inaccurately recorded.

In summary, data from Three-Day Food Diaries indicate that the majority of participants consumed adequate amounts of fat per day, but did not consume adequate calories. In addition, the nutritional intake of participants was not statistically related to adherence.

7.2. Nutritional Status

Malnutrition typically presents itself as low body weight and suboptimal growth, in the CF

population (Sinaasappel et al., 2002). Measurements of malnutrition can include BMI_p, suboptimal energy intake, HFA percentile, and WFA percentile (Lai et al., 1998; Ionescu et al., 1998, 2000; Dray et al., 2004; Steinkamp & Wiedemman, 2001). In this study, BMI_p and %IBW were determined for eight participants, since one did not self-report height and weight. It is important to note that all values were self-reported and not confirmed by anthropometric tests or clinical records. As a result, height and weight values are subject to reporting errors caused by memory or intentional misreporting.

According to the BMI_p calculations, the majority of participants were at a healthy weight (62.5%), with one being underweight and falling into the 'wasted category', and two at risk of being overweight. Since studies have found anywhere from 18-34% of participants with CF being malnourished (Lai et al., 1998), it seems unusual that there were two participants at risk of being overweight. The following factors, may contribute to this finding: participant's weight and/or height may have been inaccurately recorded, degree of pancreatic insufficiency may have been mild, lung involvement may have been mild, and/or there may have been an absence of secondary complications which may increase energy requirements. Having extra weight does not have the same consequence for those with CF as for the general population. In times of lung exacerbation, when it is common to lose weight, the extra pounds will come in handy to keep the individual's weight and nutritional status up, and thus help fight the infection.

As can be seen in Figure 8, there is quite a difference between the BMI_p and %IBW values for some of the participants. The %IBW values range from a low of 78 to a high of 125, whereas the BMI_p values range from a low of the fourth percentile to a high of the 85^{th}.One striking difference was found with participant G, at 90% of his/her ideal body weight, but only at the fourth percentile for BMI.

In attempting to explain why values for %IBW differ so much from those for BMI_p, one must go back to the underlying assumptions of the indices. Section 5.3.3. explains that each use different units and have different scales- 100% being optimal for % IBW, and the 50^{th} percentile being optimal for BMI_p (Zhang & Lai, 2004). The % IBW assumes that the ideal weight for a given height is at the same percentile ranking as the HFA (Zhang & Lai, 2004). The BMI_p assumes that the ideal weight for a given height is equal to the 50^{th} percentile value for that age (Zhang & Lai, 2004). With 100% being ideal for %IBW, four participants were at an optimal nutritional status. It is interesting to note that these same four also met or surpassed optimal values for BMI_p. A study comparing BMI_p to %IBW in the CF population found that both yield similar results for those who are within the 25^{th} to 75^{th} percentile for height (Zhang & Lai, 2004). Only two participants had a HFA value below the 25^{th} percentile- one of these being participant G, at the 10^{th} percentile, and the other being at the 20^{th}. Therefore, for these individuals, BMI_p may be a better indicator of nutritional status than %IBW.

The relationship between adherence and nutritional status (BMI_p and %IBW) was determined via two-tailed independent sample t-tests (see Table 13). For this statistical comparison, adherence was defined as those participants who were adherent to enzymes for both meals and snacks (four adherent, five non-adherent). Results indicate that there was no statistical significance between adherence level and nutritional status. However, those who were adherent tended to have a higher value for both BMI_p and %IBW.

As described in Section 7.1., the nutritional status of an individual with CF can have an impact upon the body's physiology. It has been shown that a low body weight is often one of the most consistent predictors of low bone mineral density (Bachrach et al., 1994; Turner et al.,

2001). According to recommendations from Fok et al. (2002), three participants, who had a BMI below the 25th percentile, should begin some form of aggressive nutritional therapy to help decrease their risk of developing osteoporosis or osteopenia.

When looking at the effects of nutrition on the progression of puberty, each female participant was within the weight range proposed by Frisch & Revelle to be necessary for the initiation of menstruation (Frisch, 1969; Frisch & Revell, 1971). Two of the male participants, however, may be at an increased risk of pubertal delay since their BMI is below the 25th percentile (Boas et al., 1998).

In summary, anthropometric self-reported data obtained from the Adherence Survey indicates that half of the participants were at an optimal nutritional status, as determined by %IBW and BMI_p. Those who were adherent had a higher nutritional status than those who were non-adherent, although not to a statistically significant level. Those participants who did not have an optimal nutritional status would probably benefit from a boost in caloric intake, to enhance weight gain.

7.3. Body Esteem and Disordered Eating Attitudes

Body esteem is the self-evaluation of one's body appearance, which can be different from the perceptions and opinions of others (Mendelson et al., 2001). Studies have shown that girls are more likely to rate their body appearance as negative, when compared to boys (Mendelson et al., 2001). Self-evaluations may get increasingly negative as children age and/or gain weight (Mendelson et al., 1996). Since there is a link between body-esteem, self-esteem, and self-worth (Mendelson et al., 1996), it is not surprising that recent studies show approximately 60% of

teenage girls and 25% of teenage boys have tried to lose weight in the past (Jones et al., 2001; Daee et al., 2002).

While having a negative body esteem may lead to the adoption of dieting behaviors, dieting itself can lead to negative psychological and physiological consequences, such as the adoption of more negative health behaviors, a poor body image, decreased self-esteem, loss of electrolytes, minerals, and trace metals, changes in the muscle of the heart, and adverse effects of free fatty acids on the heart muscle (Daee et al., 2002).

Research in the area of body esteem and eating disorders in the CF population is sparse. One study reported that the incidence of eating disorders in this population is low (Raymond et al., 2000). While the EAT-26 is a commonly used instrument, results in the literature are difficult to compare to this study. Some studies used different scoring mechanisms, used different cut-off levels, and were conducted on populations of various cultural backgrounds and levels of physical activity (Slater & Tiggemann, 2002; Greenleaf & McGrer, 2006; Pastore, Fisher, & Friedman, 1996). Results of the current study show a low prevalence of disordered eating attitudes, as assessed by the EAT-26. The average EAT-26 score in this study was 7.78, with one participant (out of nine), or 11%, considered at risk of developing an eating disorder. Results of this study are similar to results from a study of 140 females (mean age 20.3) without anorexia nervosa, which was found to be 9.9 (Garner et al., 1982). Results were also similar to a recent Spanish study which determined that 421 boys aged 11 to 18 years (mean age 14.3 years) had a mean EAT-26 score of 6.8 ± 7.8 (Gila, Castro, Cesena, & Toro, 2005). Rates were higher in an Ontario study which found that 13% of those aged 12 to 14, and 16% of those aged 15 to 18 (Jones et al., 2001), met or exceeded the cutoff; and in a study of undergraduate psychology

students (333 female, 138 male, aged 17-24 years) which found that 20% of females and 10% of males met or exceeded the same cutoff point of 20 (Nelson, Hughes, Katz, & Searight, 1999).

When looking at the three subscales for the EAT-26, the mean score for factor I, the dieting subscale, was 1.67 (4.28% of the highest possible score), the mean for factor II, the bulimia and food preoccupation subscale, was 1.67 (9.28% of the highest possible score), and the mean for factor III, the oral control subscale, was 4.44 (21.14% of the highest possible score). Therefore, participants exhibit a certain amount of oral control over food they do, or can, consume. Similar results were seen in a group of 221 adults with CF when compared to a group of 148 controls. In this case, a modified version of the EAT-26 was used, with a scoring mechanism from zero to five instead of zero to three used in this study (Abbott et al., 2000). Scores for factors I and II were higher in the control population than the CF population. However, scores for factor III were higher for the CF group with mean and standard deviations of 6.42 ± 4.58 for CF males, and 7.30 ± 5.07 for CF females. In addition, 6% of the CF males and 11% of the CF females reported intense pre-occupations with food, with binge eating and intended vomiting (Abbott et al.).

A high value for the oral control subscale is indicative of a large degree of self-control when it comes to food, and is associated with social pressures to gain weight (Garner & Garfinkel, 1979). This is not surprising since being told to gain/maintain weight is a common occurrence for those with CF, especially those with a low body weight. The majority of participants from the literature who score high on the EAT-26, but do not have an eating disorder, experience abnormal eating patterns which can play a psychological role in the individual (Garner & Garfmkel, 1979).

Despite the prevalence of malnutrition and a low body weight in the CF population, those

with CF may have a tendency to estimate their body weight as higher than it actually is, especially in the case of females (Truby & Paxton, 2001; Wenninger et al., 2003). The desire for females, over males, to lose weight may be a product of today's culture, which portrays the ideal woman as waif-like, but portrays the idea man as buff and muscular. Studies indicate that CF participants may be less satisfied with their body size than controls (Truby & Paxton, 2001; Abbott et al., 2000). Results from this study calculated a mean BES score of 52.67, 57.25% of the highest possible score. The majority of participants (56%) had scores between 40 and 49, with 11% having scores between 70 and 79.

Since the BES is a newly developed tool, few articles were found using the tool, except for the original studies during its development and validation (Mendelson et al.,1997). Results from a study of 1240 participants, 12 and 25 year olds, from English speaking schools, colleges, and universities in Montreal, yielded lower mean results than that of this study, with values for the subscales ranging from 2.1 to 2.9 (Mendelson et al., 1997). However, since higher scores reflect a more positive body esteem, there is probably no cause for alarm since scores in this study are higher. The mean score in this study for subscale BE weight, representing how one feels about their weight, was 17.33 (54.16% of the highest possible score), indicating some dissatisfaction with the participant's body weight. One of the typical characteristics of those with CF is a small weight. Therefore, dissatisfaction with one's weight may be due to feelings of being under- versus over-weight. This may be the case more so for males, since it has been shown that girls with CF prefer to be smaller while boys with CF prefer to be larger (Truby & Paxton, 2001; Abbott et al.).

When looking at the other two subscales, the mean score for BE appearance, which

represents how one feels about their appearance, is 22.89 (57.23% of the highest possible score), and mean BE attribution, representing how one thinks others evaluate their appearance, was 12.44 (62.2%) of highest possible score. This indicates that participants perceived others would, or do, evaluate their appearance more positively than they themselves evaluate it. This is probably not out of place in any population, as people are often their own worst critics.

The relationship between adherence, BES, and EAT-26 was investigated by two-tailed independent sample t-tests (see Table 13). For this statistical comparison, adherence was defined as those participants who were adherent to enzymes for both meals and snacks (four adherent, five non-adherent). Results indicate that there was no statistical significance between adherence level, BES, or EAT-26. However, those who were adherent tended to have a higher value for BES and a lower value for EAT-26, indicating a more positive body esteem and eating attitude in this population, versus those who were non-adherent.

In summary, one participant out of nine was at risk for developing disordered eating attitudes and/or an eating disorder. There was a low incidence of dieting, bulimia, and food preoccupation attitudes, with a higher incidence of oral control attitudes. With regards to body esteem, participants were at midline, though they feel that others would evaluate their appearance higher than they themselves evaluate it. Those who were considered adherent tend to have BES and EAT-26 scores which are more positive than those who were non-adherent, though not to a statistically significant level.

7.4. Adherence

Adherence is "an active, voluntary, collaborative involvement of the patient in a mutually

acceptable course of behavior to produce a desired preventative or therapeutic result" (Meichenbaum & Turk, 1987). Adherence levels are difficult to gauge because adherence can be viewed in different ways, defined in different ways, measured in various ways, and can change from situation to situation (Meichenbaum & Turk, 1987). Self-reports, which were used in this study, may be subject to inaccuracies, manipulation, and bias (Cluss & Epstein, 1985). Although there was a column labeled supplements on the Three-Day Food Diary, some participants may not have thought to include vitamins in this category.

In this study, adherence was measured by the Adherence Survey and the Three-Day Food Diary. Four aspects of treatment adherence were investigated: vitamins, nutritional supplements, enzymes with meals and snacks, and dietary recommendations. Perceived adherence was determined for the first three treatments by the AS, and actual adherence for the first three was determined by comparing recorded amounts in the Three-Day Food Diary with prescribed amounts recorded on the AS (Questions 3Full, 9Full, and 13).

General rates of adherence are in the vicinity of 30 to 60% for medications, and 30 to 35% for prophylactic agents (Marston, as cited in Meichenbaum & Turk, 1987). Studies specifically with the CF population found adherence rates for medication vary from around 70 to 90% (Passero et al., 1981; Gudas et al., 1991; Ievers et al., 1999). In addition, there are estimates that 50% of the pediatric CF population is non-adherent (La Greca & Schuman, as cited in Bernard & Cohen, 2004; Litt et al., 1980). In this study, the levels of actual adherence to supplements and vitamins was very poor, with all participants being considered non-adherent (all participants were prescribed vitamins and four were prescribed supplements). Levels of actual adherence was more promising with regards to enzymes taken with meals and snacks, with 44% taking less than

prescribed (considered non-adherent), 33% taking the same amount (considered adherent), and 22% taking more (considered adherent). Therefore, this study found that the rate of adherence to enzymes (55%) was similar to reported rates in the literature. However, rates for nutritional supplements and vitamins were very poor, which may be partially explained by the participants forgetting to input these two items on their diaries.

It is interesting to look at rates of perceived adherence. The largest group of participants reported that they take a full dose of vitamins (44%), enzymes with meals (67%), and enzymes with snacks (44%) every day of the week. Participants had a more realistic view of their adherence to supplements, since the four who were prescribed supplements indicated that they took a full dose of vitamins 3-4 days a week or less. Therefore, it is not that surprising that adherence levels were low for supplements.

The Adherence Survey was also designed to determine why participants are non-adherent. Questions 6, 12, and 18 provided participants a list of possible reasons as to why one might miss a treatment, as well as the opportunity to include a response not listed. With regards to vitamins, 67% chose "Sometimes I tend to forget to take them", 33% chose "I do not like to take them in front of others", 22% chose "They taste awful", and 11% chose "I do not know why I have to take vitamins", "I have problems swallowing them", "Taking the vitamins does not make me feel better", and "It takes a lot of effort or time to take them." No additional responses were given. Similar trends were seen in the literature which cite forgetfulness as a primary reason for non-adherence to vitamins (average 62%), followed by taking a lot of time or effort (participants can't be bothered to take them) (29%) (Abbott et al., 1994; Conway et al., 1996). Given the fact that one participant indicated they did not know why they were asked to take vitamins, and that

some participants may not see the long term benefits of taking vitamins, more education around vitamins may be helpful to promote adherence.

For nutritional supplements, 25% (n=1) of participants chose the following reasons for non-adherence: "Sometimes I tend to forget to take them", "The supplement/s tastes awful", "I feel that taking the supplement/s does not make my health any better", "It takes a lot of effort or time to take the nutritional supplement/s", and "I am happy at my current weight." Additional responses included: "Not in the mood", and "Very filling", both reported by one participant each. Rates of forgetfulness found in this study (25%) are similar to those found by Conway et al. (1996), at 29%. Despite the fact that no participants indicated that they do not know why they were asked to take supplements, the low level of adherence suggests that perhaps they do not see any benefits in taking supplements.

Lastly, trends for enzymes are similar to the other two treatments, with 78% choosing "I tend to forget to take them at times", 22% choosing "I do not like to take them in front of others", and 11% choosing "It takes a lot of effort or time to take them." Twenty-two percent of participants indicated that they adjust the amount of enzymes taken depending on what is eaten. This is a reasonable response and does not necessarily indicate non-adherence, since food items that are primarily sugar-based require very little, if no, enzymes compared to those that are high in fat. Rates for forgetfulness cited in the literature are a bit lower than the rate found in this study (average of 50%), while rates related to taking enzymes in front of others and taking a lot of time or effort (can't be bothered to take it or finds it interrupts social life) are similar (27% and 12%, respectively) (Abbott et al., 1994). One participant chose "I am happy at my current weight and do not want to gain weight" as a reason for non-adherence, which raises a red flag for possible

non-adherence to adjust or maintain one's weight. This was not the same participant who chose this reason for the nutritional supplements. This individual was in the risk of overweight category with respect to BMIp (87^{th} percentile), was at 125% their IBW, was in the 51-75% category for meeting adjusted EER, and was considered adherent to enzymes with meals and snacks. Therefore, this participant may not have typical CF caloric needs and may not need to gain weight. Also, perhaps this participant does not have a strong degree of pancreatic insufficiency.

As Abbott, Dodd, & Webb (1996) state, "it would be unrealistic to expect patients to comply with treatments absolutely." The literature states that treatment adherence during a chronic condition is often poorer than during an acute condition (Meichenbaum & Turk, 1987). Adherence is also negatively correlated with the duration of treatment, the number of medications or treatments being taken at one time, related side effects, an increase in age, and prophylactic treatments (Cluss & Epstein, 1985; Garcia et al., 2003; Lask, as cited in Kettler et al., 2002; Lask, 1994; Landau, 1995). Therefore, it is not surprising that adherence rates in such a small population of individuals with CF are quite low- especially for vitamins and supplements which can be considered prophylactic treatments. The majority of individuals with CF have probably been on various treatments and medications since diagnosis, and this will more than likely continue indefinitely.

Adherence to dietary recommendations was assessed by comparing daily caloric intakes from completed Three-Day Food Diaries to participant's adjusted EER's (see Section 5.3.2. for more information). Eight participants were included in this analysis since one did not provide weight and height needed to determine his/her EER. The determination of participant daily

caloric intakes, as discussed previously, probably represents a modest amount since food diaries were lacking in descriptions. As a result, certain strategies were adopted to allow for nutrient analysis (see Section 6.3.1. for more information). When calculating participant adjusted EER's, it was assumed that participants fell into the Low Active category for PAC (see Section 5.3.2.). Taking this into consideration, results indicate that one participant was considered adherent with respect to their caloric recommendations.

In summary, despite the fact that most participants were aware of why they were prescribed vitamins and nutritional supplements, none were adherent. Adherence to dietary recommendations was a little better, with one participant consuming their recommended daily calories. Such a low adherence rate may be attributed to the fact that positive effects from these prophylactic treatments are not readily seen. Rates of adherence to enzymes with meals and snacks were similar to rates in the literature, at 55% adherence. The majority of participants cite forgetfulness as their number one reason for non-adherence to vitamins, supplements, and enzymes. Given the nature of CF and its treatments, this is to be expected and was also seen in the literature.

7.5. Theory of Planned Behavior

The TPB, as explained in Section 4.1., assumes that an individual's behavior is determined by their intention to perform the behavior. With regards to this study, the key behavior which was examined was adherence to vitamins, nutritional supplements, enzymes, and diet therapy. While the investigation of the following was beyond the scope of the study, by applying the TPB, participant adherence could be influenced by these questions:

a) How does the participant view taking the treatments as recommended?

b) How does the participant think their peers view their taking the treatments as recommended?

c) Does the participant feel he/she is able to, or has control over, adhering to the treatments?

Exploring these key questions leads to the discovery of potential barriers or enablers/promoters which may influence the participant's intention to adhere. For question (a), barriers may result if the participant does not see the necessity of taking the treatment or following dietary therapy- they may not understand why it is necessary for them to take enzymes or eat according to recommendations, they may not value adherence, or they may not see its relationship to total personal health. This, in fact, was seen in the study by 11% of the participants who did not know why they had been prescribed vitamins and felt that taking vitamins did not make them feel better. As well, 25% of those who had been prescribed nutritional supplements felt that taking the supplement did not make them feel better. In contrast, if the participant values adherence and can see the bigger picture, the relationship between current adherence and future health status, they may be more likely to adhere.

For question (b), barriers may result if participants, especially females, perceive that their peers value dieting and slimness over taking medication and consuming a high fat and high calorie diet. As well, participants may perceive that their peers value inclusiveness and homogeneity in their "clique", whereas taking medication or eating foods which are different may be viewed as being unusual and may lead to exclusion from the group. The influence of peers was noted by 33% of the participants who did not like taking vitamins in front of others and 22% who did not like to take enzymes in front of others. Oppositely, the presence of

supportive friends and family, who value individuality and the maintenance of health, may foster adherence to the medical regimen. However, even in the absence of such a support group, one may not follow the opinions of others and may make independent, informed decisions to adhere to treatment.

For question (c), the participant's ability to take, and control over taking, enzymes and following dietary recommendations will impact their intentions to adhere. In Canada, most CF medications are provided through a provincial health-care program, which is a strong enabling factor (CCFF, 2003). This was reflected in the AS since no participant chose a lack of money to be a factor affecting adherence to either treatment. However, barriers such as the inability to swallow pills, being embarrassed or ashamed to take pills in front of others, or devaluing adherence, may make adherence a challenge. With regards to caloric recommendations, issues such as food insecurity may come into the picture, posing a barrier. Since the recommended caloric intake for those with CF requires consuming 20-50% more calories (Creveling et al., 1997), being food insecure may negatively affect one's ability to acquire adequate nutrition. Although supplemental feedings are available through provincial health programs, enabling individuals to achieve normal or optimal nutritional status, the brand which is provided may not be palatable (CCFF, 2003). One participant indicated that they did not like the taste of the nutritional supplement they were asked to take, and two participants indicated they did not like the taste of the vitamin/s they were asked to take.

7.6. The Health Belief Model

The Health Belief Model, as described in Section 4.2., assumes that people will adopt health

related actions if they feel a negative health condition can be avoided, and if they believe they can successfully take the action (Universiteit Twente, 2004a).

Key term III of the HBM is the perceived benefit of taking an action. As was noted in Section 7.4., adherence to prophylactic treatments is commonly lower than for treatments whose effects are readily seen. In this study, adherence to vitamins, nutritional supplements, and dietary recommendations was low. These treatments could be considered prophylactic since the benefits of adhering to them are not readily seen. However, the consequences of failing to take dietary enzymes may act to promote adherence. Adherence levels for enzymes were much higher than for other forms of treatment, at 55% adherence. If those with CF do not take enzymes, they may develop steatorrhea, diarrhea, bloating, and/or severe stomach cramps (World Health Organization Human Genetics Program & International Cystic Fibrosis Association, 1996). These unpleasant complications may provide an adequate benefit for adherence to enzymes. However, if someone has gone without enzymes before with little gastric upset, the consequences of non-adherence do not have the same impact as if he/she developed cramping or diarrhea.

Similarly, with regards to dietary therapy, one may see the slimness created by CF as a benefit and not think about the potential health risks that may occur if they are underweight and/or restricting their intake. One participant, who had been prescribed nutritional supplements, indicated that they were happy at their current weight and thus, did not always take the supplement. However, this person was at 100% of their IBW, their BMIp was in the healthy category (at 68th percentile), they were in the 79-99% category for meeting adjusted EER, and they were considered adherent to enzymes taken with meals and snacks. Therefore, perhaps it is

not vital that this individual needs to gain extra weight. Conversely, if the participant's health team is likely to prescribe enteral or supplemental feeds if they are underweight, a benefit of adhering to diet therapy could be the avoidance of such interventions (Pencharz & Durie, 1993). However, this was not investigated in the study.

Section 8: Recommendations for Future Research

8.1. Recommended Modifications for Current Study

The most notable limitation of this study was the low number of participants. Due to time and money constraints, the geographic area of recruitment was limited. While results of this study may not be typical of all Canadians with CF, the study design has the possibility to yield more fruitful results given appropriate resources.

Ideally, it would be optimal to conduct such a study Nation wide, involving all CF clinics across the country. As well, it would be beneficial for each clinic to be provided with a trained individual to interview participants, help with filling in the gaps on the Three-Day Food Diaries, and being there to answer any questions.

It is obvious that such a study would require tremendous time and human, and financial resources. For these reasons, such an endeavor was beyond the scope of this Master's thesis. However, such a study would undoubtedly reveal much useful information which could be applied in a clinical setting.

In hindsight, it would have been beneficial if physical activity level had been assessed for the participants. This would have ensured a more accurate determination of EER, instead of assuming that each participant was at the Low Active physical activity level.

It would have been ideal if the investigator could have met with the participants in person and discussed the protocol for completing the survey with the participant and his/her parent/guardian. However, due to time constraints and concerns about bacterial transmission, this was not a viable option. If finances had allowed, assistants could have been trained to deliver this information, if considered appropriate by the ethics review boards of both institutions.

8.2. Recommendations for General Research

Up to date, large scale studies assessing adherence levels to enzymes, vitamins, nutritional supplements, and dietary therapy, across many age spectrums, would be beneficial. Clinicians may take for granted that patients listen faithfully and adhere to their recommendations. However, as seen in the literature and this study, levels of adherence may be less than satisfactory, current well controlled studies would give a timely portrait of the adherence level in the CF population.

It is not enough to know if one is adherent. In order to fully address the issue of non-adherence, researchers and clinicians must be aware of reasons why one does not adhere. These reasons may be multi-layered and may involve social, socioeconomic, psychological, and physiological factors. Once it is understood why people are non-adherent, better strategies can be developed to work with the individual with CF to promote adherence.

In surveying the available literature, there were a limited number of articles investigating body esteem and eating disorders in teenagers with CF. As those with CF enter the teenage years they struggle with the same issues as healthy peers, in addition to the added burden of having a chronic illness. The presence of a chronic illness will undoubtedly make life challenging and, in efforts to fit in, teens with CF may adopt similar attitudes and behaviors as their peers. Clinicians need to be aware of the extra psychological burden CF presents to adolescents. Body image is important to all teenagers, as seen by the large number of teenagers who adopt unhealthy eating behaviors. While problematic in itself, the presence of an eating disorder in an individual with CF may be more deadly than in the healthy population. This fact strengthens the case for more research regarding the self-image and eating behaviors of those with CF.

Section 9: Relevance to the Dietetic Profession

Dietitians fall under the scope of the "helping professional"- a service-oriented professional who is committed to improving the quality of life of clients in areas in which he/she has qualified expertise, and who interacts with clients to facilitate the delivery of the service (Hodges & Vickery, 1989). The clinical CF team typically involves the contribution of a dietitian, since nutrition is such an important part of the maintenance of the individual's health.

While it is important for the dietitian to assess eating patterns, follow weight trends, and give advice on dietary matters, he/she can also play a role in assessing adherence. To try to assess adherence, dietitians could ask questions about treatment dosage and reasons for failing to take treatments. Here, "treatment" refers to enzymes, vitamins, dietary therapy, and nutritional supplements, when required. Once dietitians know the reason for non-adherence, they can work with the individual and his/her family to find solutions to overcome these problems.

Health education is another important aspect of dietetics. As the TPB states, in order for one to be adherent they have to be motivated to be adherent (Universiteit Twente, 2004). Part of this motivation exists in knowing why one needs to take the treatments they are prescribed. As children, the caregivers can assume the primary responsibility over delivering medical treatments (Pfeffer et al., 2003). With increasing age comes increasing responsibility, and the possibility of poor medical decisions and non-adherence (Landau, 1995). Given this fact, teenagers should be provided with appropriate knowledge about their disease and the nutritional implications and treatments (Ievers et al., 1999; Koocher et al., 1990).

The dietitian is in a unique position to watch for signs of low body esteem and disordered eating behaviors and attitudes. It is then that proper referrals to a mental health professional can

help ensure the individual with CF is provided the best care for their physical, as well as mental and emotional, health.

Section 10: References

Abbott, J., Conway, S., Etherington, C., Fitzjohn, J., Gee, L., Morton, A., et al. (2000). Perceived body image and eating behavior in young adults with cystic fibrosis and their healthy peers. *Journal of Behavioral Medicine, 23*(6), 501-517.

Abbott, J., Dodd, M., Bilton, D., & Webb, K.A. (1994). Treatment compliance in adults with cystic fibrosis. *Thorax, 49*, 115-119.

Abbott, J., Dodd, M., & Webb, K.A. (1995). Different perceptions of disease severity and self care between patients with cystic fibrosis, their close companions, and physician. *Thorax, 50* (7), 794-796.

Abbott, J., Dodd, M., & Webb, K.A. (December 1996). Health perceptions and treatment adherence in adults with cystic fibrosis. *Thorax, 51(*12), 1233-1238.

ACE Network. (February 25, 1999). *Eating Attitudes Test- Eating Disorder*. Retrieved June 16, 2006 from http://www.ace-network.com/eatdis/EATtest.htm

Adolescent Medicine Committee, Canadian Paediatric Society. (1994). Care of the chronically ill adolescent. *Canadian Journal of Paediatrics, 1(*4), 124-126.

Anthony, H., Paxton, S., Bines, J., & Phelan, P. (1999). Psychosocial predictors of adherence to nutritional recommendations and growth outcomes in children with cystic fibrosis. *Journal of Psychosomatic Research, 47*(6), 623-634.

Aswani, N., Taylor, C.J., McGraw, J., Pickering, M., & Rigby, A.S. (2003). Pubertal growth and development in cystic fibrosis: a retrospective review. *Acta Paediatr, 92,* 1029-1032.

Austgen, L., Bowen, R.A., & Rouge, M. (November 23, 2001). *Secretion of bile and the role of bile acids in digestion.* Colorado State University. Retrieved April 17, 2006, from http://www.vivo.colostate.edu/hbooks/pathphys/digestion/liver/bile.html

Bachrach, L.K., Loutit, C.W., Moss, R.B., & Marcus, R. (January 1994). Osteopenia in adults with cystic fibrosis. *Am J Med, 96*(1), 27-34.

Baranowski, T., & Dworkin, R., Henske, J.C., Clearman, D.R., Dunn, J.K., Nader, P.R., et al. (October 1986). The accuracy of children's self-reports of diet: Family health project. *JADA, 86*(10), 1381-85.

Beker, L.T., Ahrens, R.A., Fink, R.J., O'Brien, M.E., Davidson, K.W., Sokoll, L.J., et al. (May 1997). Effects of vitamin K supplementation on vitamin K status in cystic fibrosis patients. *J Pediatr Gastroenter Nutr, 24*(5), 512-517.

Bell, S.C., Saunders, M.A., Elborn, J.S., & Shale, D.J. (February 1996). Resting energy

expenditure and oxygen cost of breathing in patients with cystic fibrosis. *Thorax, 51(*2),126-131.

Bennett, C., & Peckham, D. (August 2002). *The genetics of cystic fibrosis*. Retrieved December 1, 2004, from http://www.cysticfibrosismedicine.com/htmldocs/CFText/genetics.htm

Bernard, R.S., & Cohen, L.L. (2004). Increasing adherence to cystic fibrosis treatment: a systematic review of behavioral techniques. *Pediatric Pulmonology, 37*, 8-16.

BioCarta: Charting Pathways of Life. (n.d.). *Cystic fibrosis transmembrane conductance regulator and beta-2 adrenergic receptor pathway*. Retrieved May 22, 2006, from http://www.biocarta.com/pathfiles/h_cftrPathway.asp

Boas, S.R., Fulton, J.A., Koehler, A.N., & Orenstein, D.M. (Sept 1998). Nutrition and pulmonary function predictors of delayed puberty in adolescent males with cystic fibrosis. *Clinical Pediatrics, 37*(9), 573-576.

BC HealthGuide Program. (2005). *Sweat test*. Retrieved September 24, 2006, from http://www.bchealthguide.org/kbase/topic/medtest/hw5781/results.htm

Brown, R.T. (2002). Risk factors for substance abuse in adolescents. *Pediatr Clin North Am, 49,* 247-255.

Bryden, K.S., Neil, A., Mayou, R.A., Peveler, R.C., et al. (Dec 1999). Eating habits, body weight, and insulin misuse. A longitudinal study of teenagers and young adults with type 1 diabetes. *Diabetes Care, 22*(12),1956-60.

Canadian Cystic Fibrosis Foundation. (2002). *Report of the Canadian cystic fibrosis patient data registry*. Retrieved August 17, 2006, from http://www.cysticfibrosis.ca/pdf/Lay%20CPDR%20ReportE.pdf

Canadian Cystic Fibrosis Foundation. (2003). *Disease information*. Retrieved September 16, 2003, from http://www.cysticfibrosis.ca/page.asp?id=1.

Canadian Cystic Fibrosis Foundation. (2004). *Drug Coverage & CF Clinics*. Retrieved October 13, 2004, from http://www.cysticfibrosis.ca/page.asp?id=192 & http://www.cysticfibrosis.ca/page.asp?id=60

Canadian Cystic Fibrosis Foundation. (2005). *Facts about CF*. Retrieved April 16, 2006, from http://www.cysticfibrosis.ca/page.asp?id=21

Canadian Diabetes Association. (November 2005). *Handy portion guide*. Retrieved August 1, 2006, from http://www.diabetes.ca/Files/plan%20your%20portions.pdf

Canadian Paediatric Society. (September 2004). Dieting in adolescence. *Paediatr Child Health, 9*(7), 487-491.

Canadian Paediatric Society (CPS), Dietitians of Canada (DC), Community Health Nurses Association of Canada (CHNAC), & The College of Family Physicians of Canada (CFPC). (March 2004). A health professional's guide to using growth charts. *Paediatr Child Health, 9*(3), 174-176.

Carlstedt-Duke, J., Bronnegard, M., & Strandvik, B. (1986). Pathological regulation of arachidonic acid release in cystic fibrosis: the putative basis defect. *Proc Natl Acad Sci USA, 83*(23), 9202-9206.

Centre for Disease Control and Prevention. (2006a). *Body Mass Index: about BMI for children and teens*. Retrieved August 2, 2006, from http://www.cdc.gov/nccdphp/dnpa/bmi/childrens_BMI/about_childrens_BMI.htm

Centre for Disease Control and Prevention. (2006b). *Body mass index: child and teen calculator.* Retrieved August 2, 2006, from http://apps.nccd.cdc.gov/dnpabmi/Calculator.aspx?CalculatorType=Metric

Chromosomal Mosaicism. (2003). *Chorionic villus sampling*. Retrieved June 12, 2006, from http://www.medgen.ubc.ca/wrobinson/mosaic/cvs.htm

Cluss, P.A., & Epstein, L.H. (1985). The measurement of medical compliance in the treatment of disease. In: Karoly P, ed. *Measurement strategies in health psychology*. New York: John Wiley and Sons. pp. 403–32.

Collins, C.E, O'Loughlin, E.V., & Henry, R. (March 1998). Discrepancies between males and females with cystic fibrosis in dietary intake and pancreatic enzyme use. *Journal of Pediatric Gastroenterology and Nutrition, 26*, 258-262.

Conway, S.P. (2001). Impact of lung inflammation on bone metabolism in adolescents with cystic fibrosis. *Paediatric Respiratory Reviews, 2*, 24-331.

Conway, S.P., Morton, A.M., Oldroyd, B., et al. (2000). Osteoporosis and osteopenia in adults and adolescents with cystic fibrosis: prevalence and associated factors. *Thorax, 55(*9), 798-804.

Conway, S.P., Pond, M.N., Hamnett, T., & Watson, A. (Jan 1996). Compliance with treatment in adult patients with cystic fibrosis. *Thorax, 51*(1), 29-33.

Coppenhaver, D., Kneppers, F., Schidlow, D., Bee, D., Isenburg, J.N., Barnett, D.R., et al. (1981). Serum concentrations of vitamin D-binding protein (group-specific component) in cystic fibrosis. *Human Genetics, 57*, 399-403.

Corey, M., McLaughlin, F.J., Williams, M., & Levinson, H. (1987). A comparison of survival, growth, and pulmonary function in patients with cystic fibrosis in Boston and Toronto. *J Clin Epidemiol, 41(*6), 583-591.

Creveling, S., Light, M., Gardner, P., & Greene, L. (Oct 1997). Cystic fibrosis, nutrition, and the health care team. *JADA*, *10* (Suppl. 2), S186-191.

Crozier, D.N. (1974). Cystic fibrosis- a not so fatal disease. *Pediatric Clinics of North America, 21*(4), 935-950.

Daee, A., Robinson, P., Lawson, M., Turpin, J.A., Gregory, B., & Tobias, J.D. (2002). Psychologic and physiologic effects of dieting in adolescents. *South Med J, 95*(9), 1032-1041.

DeLambo, K.E., Ievers-Landis, C.E., Drotar, D., & Quittner, A.L. (2004). Association of observed family relationship quality and problem solving skills with treatment adherence in older children and adolescents with cystic fibrosis. *Journal of Pediatric Psychology, 29*(5), 343-353.

De Vriese, S., Robberecht, E., De Gussem, J., & Christophe, A. (June 1999). FEV1 correlates with plasma vitamin E in young cystic fibrosis patients: an argument for early administration? *Netherlands J Med, 54* (Suppl. 2), S61.

Dietitians of Canada, Canadian Paediatric Society, The College of Family Physicians of Canada, & The Community Health Nurses Association of Canada. (2004). *The use of growth charts for assessing and monitoring growth in Canadian infants and children.* Retrieved October 2004, from http://www.dietitians.ca/news/highlights_positions.html

Dietz, W.H. Jr., & Hartung, R. (July 1985). Changes in height velocity of obese preadolescents during weight reduction. *Am J Dis Child, 139*, 705-7.

Dray, X., Kanaan, R., Bienvenu, T., Desmazes-Dufeu, N., Dusser, D., Marteau, P., et al. (2004). Malnutrition in adults with cystic fibrosis. *Eur J Clin Nutr,* 1-3.

Drotar, D., & Ievers, C. (Aug 1994). Age differences in parent and child responsibilities for the management of cystic fibrosis and insulin-dependent diabetes mellitus. *Developmental and Behavioral Pediatrics,15*(4), 265-272.

Durie, P.R., Kalnins, D., & Ellis, L. (1998). Uses and abuses of enzyme therapy in cystic fibrosis. *J Roy Soc Med, 91*(Suppl. 34), 2-13.

Durie, R.R., & Pencharz, P.B. (1989). A rational approach to the nutritional care of patients with cystic fibrosis. *J R Soc Med, 82*(Suppl. 16), 11-20.

Easley, D, Krebs, N, Jefferson, M., Miller, L., Erskine, J., Accurso, F., et al. (February 1998). Effect of pancreatic enzymes on zinc absorption in cystic fibrosis. *J Pediatr Gastroenterol Nutr, 26(*2), 136-139.

Elborn, J.S., & Bell, S.C. (Oct 1996). Nutrition and survival in cystic fibrosis. *Thorax*, *51*, 971-972.

Elborn, J.S., Cordon, S.M., Western, P., MacDonald, I.A., & Shale, D.J. (1993). Tumor necrosis factor alpha, resting energy expenditure and cachexia in cystic fibrosis. *Clin Sci, 85*, 563-68.

Eliot, A.O., & Baker, C.W. (Fall 2001). Eating disordered adolescent males. *Adolescence*, *36*(143), 535-543.

Elkin, S.L., Fairney, A., Burnett, S., Kemp, M., Kyd, P., Burgess, J., et al. (May 2001). Vertebral deformities and low bone mineral density in adults with cystic fibrosis: a cross sectional study. *Osteoporosis Int, 12(*5), 366-372.

Feranchak, A.P., Sontag, M.K., Wagener, J.S., Hammond, K.B., Accurso, F.J., & Sokol, R.J. (Nov 1999). Prospective, long term study of fat-soluble vitamin status in children with cystic fibrosis. *J Pediatr, 135(*5), 601-610.

Fishbein, M., & Ajzen, I. (January 2005). Theory-based behavior change interventions: comments on Hobbis and Sutton. *J Heal Psychol, 10*(1), 27-31.

Fok, J., Brown, N.E., Zuberbuhler, P., Tabak, J., & Tom, M. (Winter 2002). Low bone mineral density in cystic fibrosis patients. *Canadian Journal of Dietetic Practice and Research, 63*(4), 192-197.

Frisch, R. (1976). Fatness of girls from menarche to age 18 years with a nomogram. *Hum Biol, 48*(2*),* 353-359.

Frisch, R., & Revelle, R. (1971). Height and weight at menarche and a hypothesis of menarche. *Arc Dis Child, 46,* 695-701.

Garcia, R., Schooley, R.T., & Badaro, R. (2003). An adherence trilogy is essential for long-term HAART success. *The Brazilian Journal of Infectious Diseases, 7*(5), 307-314.

Garner, D.M., & Garfinkel, P.E. (1979). The eating attitudes test: an index of the symptoms of anorexia nervosa. *Psychological Medicine, 9,* 273-279.

Garner, D.M., Olmsted, M.P., Bohr, Y., & Garfinkel, P.E. (1982). The eating attitudes test: psychometric features and clinical correlates. *Psychological Medicine*, *12*, 871-878.

Gaskin, K.J., Durie, P.R., Corey, M., Wei, P., & Forstner, G.G. (1982). Evidence for a primary

defect of pancreatic HCO3- secretion in cystic fibrosis. *Pediatr Res, 16*, 554-557.

Gene Gateway- Exploring Genes and Genetic Diseases (n.d.). *Cystic Fibrosis Gene*. U.S. Department of Energy Human Genome Project. Retrieved May 22, 2006, from http://genome.gsc.riken.go.jp/hgmis/posters/chromosome/cftr.html

Gila, A., Castro, J., Cesena, J., & Toro, J. (2005). Anorexia nervosa in male adolescents: body image, eating attitudes, and psychological traits. *J Adol Health, 36,* 221-226.

Greenleaf, C., & McGreer, R. (2006). Disordered eating attitudes and self-objectification among physically active and sedentary female college students. *J Psych, 140*(3), 187-198.

Gudas, L.J., Koocher, G.P., & Wypij, D. (1991). Perceptions of medical compliance in children and adolescents with cystic fibrosis. *J Dev and Beh Pediat, 12*(4), 236-242.

Health Canada. (2005a). *Food and nutrition: Canada's Food Guide to Healthy Eating.* Retrieved August 1, 2006, from http://www.hc-sc.gc.ca/fn-an/food-guide-aliment/fg_rainbow-arc_en_ciel_ga_e.html

Health Canada. (2005b). *Food and nutrition: Dietary Reference Intakes tables.* Retrieved August 1, 2006, from http://www.hc-sc.gc.ca/fn-an/nutrition/reference/table/index_e.html#eeer

Hodges, P.A.M., & Vickery, C.E. (1989). *Effective counseling strategies for dietary management*. Maryland, Aspen Publishers Inc.

Holliday, K.E., Allen, J.R., Waters, D.L., Gruca, M.A., Thomspon, S.M., & Gaskin, K.J. (January 1991). Growth of human milk-fed and formula-fed infants with cystic fibrosis, *J Pediat, 118*(1), 77-79.

Ievers, C.E., Brown, R.T., Drotar, D., Caplan, D., Pishevar, B.S., & Lambert, R.G. (Oct 1999). Knowledge of physician prescriptions and adherence to treatment among children with cystic fibrosis and their mothers. *Developmental and Behavioral Pediatrics*, *20*(5), 335-343.

Ionescu, A.A., Chatham, K., Davies, C.A., Nixon, L.S., Enright, S., & Shale, D.J. (1998). Inspiratory muscle function and body composition in cystic fibrosis. *Am J Respir Crit Care Med, 158(*4), 1271-1276.

Ionescu, A.A., Nixon, L.S., Buss, D., Routledge, P.A., & Shale, D.J. (1999). Urinary excretion of pseudouridine is increased in adults with cystic fibrosis and correlates with bone resorption markers. *Pediatr Pulmonol, S19*, 296.

Ionescu, A.A., Nixon, L.S., Evans, W.D., Stone, M.D., Lewis-Jenkins, V., Chatham, K., et al. (Sept 2000). Bone density and turnover, body composition and inflammatory status in

cystic fibrosis. *Am J Respir Crit Care Med, 162(*3), 789-794.

Ionescu, A.A., Nixon, L.S., Luzio, S., Lewis-Jenkins, V., Evans, W.D., Stone, M.D., et al. (2002). Pulmonary function, body composition, and protein catabolism in adults with cystic fibrosis. *Am J Respir Crit Care Med, 165*, 495-500.

Johannesson, M., Carlson, M., Brucefors, A.B., & Hjelte, L. (1998). Cystic fibrosis through a female perspective: psychosocial issues and information concerning puberty and motherhood. *Patient Education and Counseling, 34*, 115-123.

Johannesson, M., Gottlieb, C., & Hjelte, L. (Jan 1997). Delayed puberty in girls with cystic fibrosis despite good clinical status. *Pediatrics, 99*(1), 29-33.

Jones, J.M., Bennett, S., Olmsted, M.P., Lawson, M.L., Rodin, G. (2001). Disordered eating attitudes and behaviors in teenaged girls: a school based study. *Canadian Medical Association Journal, 165*(5), 547-552.

Kettler, L. (2004). *Treatment adherence in adults with cystic fibrosis*. Retrieved November 30, 2004, from http://www.cfww.org/pub/edition_2/treatment_adherence_in_adults

Kettler, L.J., Sawyer, S.M., Winefield, H.R., & Greville, H.W. (May 2002). Determinants of adherence in adults with cystic fibrosis. *Thorax, 57*(5), 459-464.

Konstan, M.W., Butler, S.M., Wohl, M.B., Stoddard, M., Matousek, R., Wagener, J.S., et al. (2003). Growth and nutritional indexes in early life predict pulmonary function in cystic fibrosis. *Journal of Pediatrics, 142*, 624-630.

Koocher, G., McGrath, M., & Gudas, L. (1990). Typologies of non adherence in cystic fibrosis. *Develop Behav Paediatr, 11*(6), 353-358.

Koslowsky, M., Scheinberg, Z., Bleich, A., Mark, M., Apter, A., Danon, Y., et al. (1992). The factor structure and criterion validity of the short form of the eating attitudes test. *J Personality Assess, 58*(1), 27-35.

Krebs, N.F., Sontag, M., Accurso, F.J., & Hambidge, K.M. (Dec 1998). Low plasma zinc concentrations in young infants with cystic fibrosis. *J Pediatr, 133(*6), 761-764.

Lai, H.C., Kosorok, M.R., Sondel, S.A., Chen, S.T., FitzSimmons, S.C., Green, C.G. et al. (March 1998). Growth status in children with cystic fibrosis based on the National Cystic Fibrosis Patient Registry data: Evaluation of various criteria used to identify malnutrition. *J Pedia, 3*(132), 478-485.

Lancellotti, L., D'Orazio, C., Mastella, G., Mazzi, G., & Lippi, U. (March 11, 1996). Deficiency of vitamins E and A in cystic fibrosis is independent of pancreatic function and current enzyme and vitamin supplementation. *Eur J Pediatr, 155(*4), 281-285.

Landon, C., Rosenfield, R.G., Northcraft, G., & Lewiston, N. (1980). Self-image of adolescents with cystic fibrosis. *J Youth Adolesc, 9*(6), 521-526.

Landau, L.L. (Oct 1995). Cystic fibrosis: transition from pediatric to adult physician's care. *Thorax, 50*(10), 1031-1032.

Lask, B. (1994). Non-adherence to treatment in cystic fibrosis. *Journal of the Royal Society of Medicine, S21*(87), 25-27.

Legris, G.J., Dearborn, D., Stern, R.C., Geiss, C.L., Hopfer, U., Douglas, J. G., & Doershuk, C.F. (January 1998). Sodium space and intravascular volume: dietary sodium effects in cystic fibrosis and healthy adolescent subjects. *Pediatrics, 101*(1), 48-56.

Litt, I.F., Cuskey, W.R., & Rudd, S. (April 1980). Identifying adolescents at risk for noncompliance with contraceptive therapy. *J Pediatr, 96*(4), 742-5.

Littlewood, J. (April 2002). *The historical development of nutritional and dietetic management of cystic fibrosis*. Retrieved September 20, 2006, from http://www.cysticfibrosismedicine.com/public/articles_text.asp?id=108

Livingstone, M.B.E., & Robson, P.J. (2000). Measurement of dietary intake in children. *Proceedings of the Nutrition Society, 59,* 279-293.

Marin, V.B., Velandia, S., Hunter, B., Gattas, V., Fielbaum, O., Herrera, O., et al. (2004). Energy expenditure, nutrition status, and body composition in children with cystic fibrosis. *Nutrition, 20*(2), 181-186.

Medicine.net. (2006). Retrieved June 12, 2006, from http://www.medterms.com

Mendelson, B.K., Mendelson, M.J., & White, D.R. (2001). Body-esteem scale for adolescents and adults. *Journal of Personality Assessment, 76*(1), 90-106.

Mendelson, B.K., White, D.R., & Mendelson, M.J. (1997). Manual for the body-esteem scale for adolescents and adults. *Concordia University Research Bulletin, 16*(2).

Mendelson, B.K., White, D.R., & Mendelson, M.J. (1996). Self-esteem and body esteem: effects of gender, age, and weight. *J Applied Dev Psychology, 17*, 321-346.

Meichenbaum, D., & Turk, D.C. (1987). Treatment adherence: terminology, incidence and conceptualization. In: *Facilitating treatment adherence*. New York: Plenum Press. pp. 19–39.

Merck Source. (2006). *Dorland's Medical Dictionary*. Retrieved June 12, 2006, from http://www.mercksource.com/pp/us/cns/cns_hl_dorlands.jspzQzpgzEzzSzppdocszSzuszSzcommonzSzdorlandszSzdorlandzSzdmd-a-b-000zPzhtm

Mulberg, A.E., Resta, L.P., Wiedner, E.B., Altschuler, S.M., Jefferson, D.M., & Broussard, D.L. (1995). Expression and localization of the cystic fibrosis transmembrane conductance regulator mRNA and its protein in rat brain. *J Clin Invest, 96*(1), 646-652.

Murphy, J.L., Wooton, S.A., Bond, S.A., & Jackson, A.A. (1991). Energy content of stools in normal healthy controls and patients with cystic fibrosis. *Arch Dis Child, 66*, 495-506.

National Library of Medicine. (2006). *Sweat electrolytes*. Retrieved September 20, 2006, from http://www.nlm.nih.gov/medlineplus/ency/imagepages/9955.htm

Nelson, W.L., Hughes, H.M., Katz, B., & Searight, H.R. (1999). Anorexic eating attitudes and behaviors of male and female college students. *Adoles, 34*(135), 621-633.

Neumark-Sztainer, D., Story, M., Dixon, L.B., & Murray, D.M. (June 1998). Adolescents engaging in unhealthy weight control behaviors: are they at risk for other health-compromising behaviors? *Am J Public Health, 88*(6), 952-959.

O'Rawe, A., McIntosh, I., Dodge, J.A., Brock, D.J., Redmond, A.O., Ward, R., et al. (1992). Increased energy expenditure in cystic fibrosis is associated with specific mutations. *Clin Sci, 82,* 71-76.

Orenstein, D.M., Winnie, G.B., & Altman, H. (2002). Cystic fibrosis: a 2002 update. *Journal of Pediatrics, 140,* 156-164.

Ortiz Zein, C., & Lindor, K.D. (January 2000). Chronic cholestatic liver disease in adults. *Best Practice of Medicine.* Retrieved June 5, 2006, from http://merck.micromedex.com/index.asp?page=bpm_report&article_id=CPM02HP381&section=report&ss=1

Passero, M.A., Remor, B., Salomon, J. (April 1981). Patient-reported compliance with cystic fibrosis therapy. *Clinical Pediatrics, 20*(4), 264-268.

Pastore, D.R., Fisher, M., & Friedman, S.B. (1996). Abnormalities in weight status, eating attitudes, and eating behaviors among urban high school students. *J Adol Health, 18*, 312-319.

Peckham, D., & Littlewood, J. (July 2004). *The sweat test*. Retrieved December 1, 2004, from http://www.cysticfibrosismedicine.com/htmldocs/CFText/sweat.htm

Peckham, D., & Morton, A. (July 2003). *Treatment of distal intestinal obstruction syndrome*. Retrieved November 21, 2004, from http://www.cysticfibrosismedicine.com/htmldocs/CFText/dios.htm

Pencharz, P.B., & Durie, P.R. (1993). Nutritional management of cystic fibrosis. *Annual Reviews of Nutrition*, *13*, 111-136.

Peters, S.A., & Rolles, C.J. (1993). Vitamin therapy in cystic fibrosis-a review and rationale. *J Clin Pharm Ther, 18,* 33-38.

Peters, S.A., & Kelly, F.J. (1996). Vitamin E supplementation in cystic fibrosis. *J Pediatric Gastroenterology and Nutrition, 22,* 341-345.

Pfeffer, P.E., Pfeffer, J.M., & Hodson, M.E. (2003). The psychosocial and psychiatric side of cystic fibrosis in adolescents and adults. *Journal of Cystic Fibrosis, 2*, 61-68.

Prentice, A.M. (November 1998). Body mass index standards for children. *British Medical Journal, 317(*7170), 1401-1402.

Quanjer, P.H., et al. (n.d.). *Lung function indices*. Retrieved June 12, 2006, from http://www.spirxpert.com/indices.htm

Rashid, M., Durie, P., Andrew, M., et al. (September 1999). Prevalence of vitamin K deficiency in cystic fibrosis. *Acta Pediatr*, *70*(3), 378-382.

Raymond, N.C., Chang, P., Crow, S.J., Mitchell, J.E., Dieperink, B.S., Beck, M.M., et al. (2000). Eating disorders in patients with cystic fibrosis. *Journal of Adolescence*, *23*, 359-363.

Reiter, E.O., Stern, R.C., & Root, A.W. (1982). The reproductive endocrine system in cystic fibrosis 2: changes in gonadotropins and sex steroids following LHRH. *Clin Endocrin, 16*, 127-37.

River Centre Clinic. (n.d.). *The Eating Attitudes Test (EAT-26)*. Retrieved October 31, 2003, from http://river-centre.org/OrderEAT26.html or, http://river-entre.org/pretest.html

Roulet, M., Franscarolo, P., Rappaz, I., & Pilet, M. (November 20, 1997). Essential fatty acid deficiency in well nourished young cystic fibrosis patients. *Eur J Pediatr, 156*(12), 952-956.

Sawyer, S.M., Rosier, M.J., Phelan, P.D., & Bowes, G. (1995). The self-image of adolescents with cystic fibrosis. *Journal of Adolescent Health*, *16*, 204-208.

Schoni, M.H., & Casaulta-Aebischer, C. (2000). Nutrition and lung function in cystic fibrosis patients: review. *Clinical Nutrition, 19*(2), 79-85.

Schur, E.A., Sanders, M., & Steiner, H. (January 2000). Body dissatisfaction and dieting in young children. *Int J Eat Dis, 27*(1), 74-82.

Selzer, R., Caust, J., Hibbert, M., Bowes, G., & Patton, G. (July 1996). The association between secondary amenorrhea and common eating disordered weight control practices in an adolescent population. *J Adol Health, 19*(1), 56-61.

Simmons, R., Corey, M., Cowen, L., Keenan, N., Robertson, J., & Lewinson, H. (March-April 1985). Emotional adjustment of early adolescents with cystic fibrosis. *Psychosom Med, 47*(2), 111-122.

Sinaasappel, M., Stern, M., Littlewood, J., Wolfe, S., Steinkamp, G., Heijerman, H.G.M., et al. (2002).Nutrition in patients with cystic fibrosis: a European consensus. *Journal of Cystic Fibrosis, 1,* 51-75.

Slater, A., & Tiggemann, M. (May 2002). A test of objectification theory in adolescent girls. *Sex Roles, 46*(9-10), 343-349.

Smolak, L., & Levine, M.P. (1994). Psychometric properties of the children's eating attitudes test. *International Journal of Eating Disorders*, *16*(3), 275-282.

Smith, S.D., Rosen, D., Trueworthy, R.C., & Lowman, J.T. (January 1979). A reliable method for evaluating drug compliance in children with cancer. *Cancer, 43*, 169-73.

Steinkamp, G., Demmelmair, H., Ruhl-Bagheri, I., von der Hardt, H., & Koletzko, B. (Oct 2000). Energy supplements rich in linoleic acid improve body weight and essential fatty acid status of cystic fibrosis patients. *J Pediatr Gastroenterol Nutr, 31(*4), 418-423.

Steinkamp, G., & Wiedemann, B. (2002). Relationship between nutritional status and lung function in cystic fibrosis: cross sectional and longitudinal analyses from the German CF quality assurance (CFQA) project. *Thorax, 57*, 596-601.

Strandvik, B., Gronowitz, E., Enlund, F., Martinsson, T., & Wahlstrom, J. (Nov 2001). Essential fatty acid deficiency in relation to genotype in patients with cystic fibrosis. *J Pediatr, 139(*5), 650-655.

Stark, L., Mulvihill, M.M., Jelalian, E., Bowen, A.M., Powers, S.W., Tao, S., et al. (May 1997). Descriptive analysis of eating behavior in school-age children with cystic fibrosis and healthy control children. *Pediatrics, 99*(5), 665-671.

Tanner, J.M. (1962). *Growth at adolescence*. Oxford: Blackwell Scientific Publications. pp. 28-39.

Thomas, P.S., Bellamie, G., & Geddes, D. (November 4 1995). Malabsorption of vitamin E in cystic fibrosis improved after ursodeoxycholic acid. *Lancet, 346* (4), 1230-1.

Tizzano, E.F., Silver, M.M., Chitayat, D., Benichou, J.C., & Buchwald, M. (May 1994). Different cellular expression of cystic fibrosis transmembrane conductance regulator in human reproductive tissues. Clues for the infertility of patients with cystic fibrosis. *Am J Pathol, 144*(5), 906-14.

Truby, H., & Paxton, S.J. (June 2001). Body image and dieting behavior in cystic fibrosis. *Pediatrics*, *107*(6), e92.

Turner, J.M., Bulsara, M.K., McDermott, B.M., Byrne, G.C., Prince, R.L., & Forbes, D.A. (November 2001). Predictors of low bone density in young adolescent females with anorexia nervosa and other dieting disorders. *Int J Eat Disord, 30*(3), 245-251.

UK Cystic Fibrosis Trust. (2002). *Nutrition: eating well with cystic fibrosis. A guide for children and parents.* Retrieved October 2004, from http://www.cftrust.org.uk/scope/documentlibrary/CFTRust/Eating%20well.pdf

UK Cystic Fibrosis Trust Nutrition Working Group. (April 2002). *Nutritional management of cystic fibrosis.* Retrieved October 2004, from http://www.cftrust.org.uk/scope/documentlibrary/CFTRust/Nutritional%20Management%20of%20Cystic%20Fibrosis.pdf

Universiteit Twente. *Health Belief Model.* Retrieved October 8, 2004a, from http://www.tcw.utwente.nl/theorieenoverzicht/Theory%20Clusters/Health%20Communication/Health_Belief_Model.doc/index.html

Universiteit Twente. *Theory of Planned Behavior/Reasoned Action*. Retrieved October 8, 2004b, from http://www.tcw.utwente.nl/theorieenoverzicht/Theory%20Clusters/Health%20Communication/theory_planned_behavior.doc/index.html

USDA Nutrient Data Products and Services. (2006). *USDA National Nutrient Database SR-18.* Retrieved July 10, 2006 from http://www.ars.usda.gov/Services/docs.htm?docid=5720

Walkowiak, J., & Przyslawski, J. (2003). Five-year prospective analysis of dietary intake and clinical status in malnourished cystic fibrosis patients. *Journal of Human Nutrition and Dietetics, 16*, 225-231.

Walters, S. (2001). Sex differences in weight perception and nutritional behavior in adults with cystic fibrosis. *J Hum Nutr Dietet, 14*, 83-91.

Wenninger, K., Weiss, C., Wahn, U., & Staab, D. (Feb 2003). Body image in cystic fibrosis-development of a brief diagnostic scale. *Journal of Behavioral Medicine, 26*(1), 81-94.

Whitney, E.N., & Rolfes, S.R. (1999). Understanding Nutrition, 8th edition. Belmont, California: Wadsworth Publishing Company.

Wikipedia. (2006). Retrieved on June 12, 2006, from: http://en.wikipedia.org/wiki/Main_Page

Winklhofer-Roob, B.M., Shmerling, D.H., Schimek, M.G., & Tuchschmid, P.E. (1992). Short term changes in erythrocyte alpha tocopherol content of vitamin E- deficient patients with cystic fibrosis. *Am J Clin Nutr, 55*, 100-103.

Wolfe, S., Tremlett, K., White, H., & Littlewood, J. (January 2001). *Pancreatic enzymes*.

Retrieved on December 1, 2004, from: http://www.cysticfibrosismedicine.com/htmldocs/CFText/enzymes.htm

World Health Organization Human Genetics Program & International Cystic Fibrosis Association. (1996). *Manual for cystic fibrosis patients and their parents.* Retrieved May 4, 2006, from http://www.cfww.org/WHO/manual%20for%20CF%20patients%20and%20their%20parents%2096.6.pdf

World Health Organization Scientific Group. (2003). *WHO Technical Report Series. Prevention and management of osteoporosis.* Retrieved June 2, 2006, from http://whqlibdoc.who.int/trs/WHO_TRS_921.pdf

Wright, A.I., & Morgan, W.J. (1990). On the creation of 'problem' patients. *Soc Sci Med, 30*(9), 951-9.

Yi, M.S., Britto, M.T., Wilmott, R.W., Kotagal, U.R., Eckman, R.H., Nielson, D.W., et al. (February 2003). Health values of adolescents with cystic fibrosis. *J Pediat*, 133-140.

Zemel, B.S., Jawad, A.F., FitzSimmons, S., & Stallings, V.A. (Sept 2000). Longitudinal relationship among growth, nutritional status, and pulmonary function in children with cystic fibrosis: analysis of the Cystic Fibrosis Foundation National CF Patient Registry. *Journal of Pediatrics, 137*(3), 74-380.

Zhang, Z., & Lai, H.J. (2004). Comparison of the use of body mass index percentiles and percentage of ideal body weight to screen for malnutrition in children with cystic fibrosis. *Am J Clin Nutr, 80,* 982-991.

Appendix A

IWK Cover Letter

Dear Participant,

I would like to introduce Colleen Faulkner who is responsible for a study at the IWK as one of the two organizations (IWK and the Janeway) participating in her research as part of her Masters Thesis at Mount Saint Vincent University. Colleen, who has cystic fibrosis (CF), is studying how teens with CF feel about their bodies, to find out about their eating habits, and to find out how often they miss doses of medication and/or nutritional supplements. As someone with CF you are being invited to take part in this study.

Included in this package are the following items:

- ✓ This cover letter;
- ✓ Instructions on how to complete the surveys;
- ✓ Study Debriefing Form (DF) (tells you about the study);
- ✓ The following surveys:
  - The Adherence Survey (AS);
  - The Body Esteem Scale (BES);
  - The Eating Attitudes Test (EAT);
  - A Three-Day Food Diary Form (FD); and
- ✓ A Communication of Results Card (CR), and
- ✓ A self-addressed stamped envelope.

I would like you to read the Debriefing Form with a parent/guardian. Taking part in the study is up to you and your parent/guardian. You are free to quit at ANY time.

If you want to take part in the study, please fill out each of the surveys. If any of the questions upset you, or if you do not want to answer a question, feel free to leave it out. When you have filled out all of the surveys, please mail them back in the self-addressed stamped envelope. You DO NOT need to put your name or address on anything, including the envelope. You DO NOT need to return this cover letter and the Debriefing Form.

If you have ANY questions about the study or the surveys, you can contact me, Colleen, or Janette Taper, who is Colleen's thesis advisor. When calling Colleen, you may call collect. You do not have to give your name, just say you are in the CF study.

I can be reached at:
- ✓ IWK Health Centre
  5850/5980 University Ave
  P.O. Box 3070
  Halifax, NS B3J 3G9
  (902) 470-7875

Colleen can be reached at:
- ✓ 2060 Quingate Place
  Apt 1115
  Halifax, NS B3L 4P7
  (902) 442-2882/ (709) 468-7943
  60Hcolleen.faulkner@msvu.ca

Janette can be reached at:
- ✓ Room 311, Evaristus Hall
  MSVU, Halifax, NS
  B3M 2J6
  (902) 457-6256 (w)
  59Hjanette.taper@msvu.ca

Sincerely,
Sheri Rose, B.Sc., P.Dt. Colleen Faulkner

Appendix B

Janeway Cover Letter

Dear Participant,

I would like to introduce Colleen Faulkner who is responsible for a study at the Janeway as one of the two organizations (Janeway and the IWK) participating in her research as part of her Masters Thesis at Mount Saint Vincent University. Colleen, who has cystic fibrosis (CF), is studying how teens with CF feel about their bodies, to find out about their eating habits, and to find out how often they miss doses of medication and/or nutritional supplements. As someone with CF you are being invited to take part in this study.

Included in this package are the following items:

- ✓ This cover letter;
- ✓ Instructions on how to complete the surveys;
- ✓ Study Debriefing Form (DF) (tells you about the study);
- ✓ The following surveys:
  - The Adherence Survey (AS);
  - The Body Esteem Scale (BES);
  - The Eating Attitudes Test (EAT);
  - A Three-Day Food Diary Form (FD); and
- ✓ A Communication of Results Card (CR), and
- ✓ A self-addressed stamped envelope.

I would like you to read the Debriefing Form with a parent/guardian. Taking part in the study is up to you and your parent/guardian. You are free to quit at ANY time.

If you want to take part in the study, please fill out each of the surveys. If any of the questions upset you, or if you do not want to answer a question, feel free to leave it out. When you have filled out all of the surveys, please mail them back in the self-addressed stamped envelope. You DO NOT need to put your name or address on anything, including the envelope. You DO NOT need to return this cover letter and the Debriefing Form.

If you have ANY questions about the study or the surveys, you can contact me, Colleen, or Janette Taper, who is Colleen's thesis advisor. When calling Colleen, you may call collect. You do not have to give your name, just say you are in the CF study.

I can be reached at:
Betty Sheppard
Room 2J732, HSC
300 Prince Phillip Dr.
St. John's, NL A1B 3V6
(709) 777-4389
66HBetty.Sheppard@hccsj.nl.ca

Colleen can be reached at:
✓ 2060 Quingate Place
Apt 1115
Halifax, NS B3L 4P7
(902) 442-2882/ (709) 468-7943
65Hcolleen.faulkner@msvu.ca

Janette can be reached at:
✓ Room 311, Evaristus Hall
MSVU, Halifax, NS
B3M 2J6
(902) 457-6256 (w)
64Hjanette.taper@msvu.ca

Sincerely,
Betty Sheppard, R.N. Colleen Faulkner

Appendix C

Survey Instructions

The Adherence Survey:

- ✓ This survey is 7 pages long
- ✓ This survey is called AS for short
- ✓ This survey has pink highlights
- ✓ There are 4 sections to the survey:
  - Vitamins
  - Nutritional Supplements
  - Enzymes
  - Demographic Information
- ✓ Please read each section and question carefully
- ✓ Please complete all sections as well as you can
- ✓ Examples are sometimes given, in black color font, to help you understand the questions
- ✓ If you have any questions please contact Colleen at (902) 442-2882 or (709) 468-7943 (collect) or via email at colleen.faulkner@msvu.ca.

The Body Esteem Scale for Adolescents and Adults:

- ✓ This survey is 1 page long
- ✓ This survey is called BES for short
- ✓ This survey has blue highlights
- ✓ Please read the statements carefully
- ✓ Please think about how often you agree with the given statements, ranging from "never" to "always".
- ✓ Please check a box beside your response for all statements, unless one upsets you or you would prefer not to answer
- ✓ If you have any questions, please contact Colleen at (902) 442-2882 or (709) 468-7943 (collect) or via email at colleen.faulkner@msvu.ca.

The Eating Attitudes Test-26:

- ✓ This survey is 2 pages long
- ✓ This survey is called EAT for short
- ✓ This survey has yellow highlights
- ✓ Please read the statements carefully
- ✓ Please think about how often you agree with the given statements, ranging from "always" to "never".
- ✓ Please check a box beside your response for all statements, unless one upsets you or you would prefer not to answer
- ✓ If you have any questions, please contact Colleen at (902) 442-2882 or (709) 468-7943 (collect) or via email at colleen.faulkner@msvu.ca.

The Three-Day Food Diary:

- ✓ The Three-Day Food Diary is 6 pages long (2 pages per day)
- ✓ This survey has orange highlights

- ✓ The diary is called FD for short
- ✓ Please record all food and drink eaten, except for water, for **3 days** in a row.
- ✓ It is better to record during the meal or snack
- ✓ It is better to include one weekend day: example record on Sunday, Monday, and Tuesday
- ✓ Describe foods you eat **in detail:**
    - **Give the amount** of food you ate by using amounts like: cups, teaspoons, tablespoons, or recording the size of the food
    - **Give brand names** if possible
    - Write down **how the food was prepared-** ways of preparing food include boiling, roasting, baking, broiling, frying, or steaming
    - For **mixed dishes** (casseroles, stews, cookies, cakes, pies, etc.) try to include the recipe on a new sheet-don't forget to write down how much of each ingredient was used, how much the recipe made, and how much of the recipe you ate
    - If eating out, **name the restaurant/chain** and record foods eaten with portion size.
    - Write down if fruits and vegetables are fresh, canned, cooked, or frozen
    - Don't forget about extra foods served with cereals or desserts like sugar, cream, butter, etc.
- ✓ Don't forget to write down the name and amount of **any vitamins and/or nutritional supplements** if taken.
- ✓ Don't forget to write down the amount of **enzymes taken** with each meal and snack.
- ✓ Examples:
    - For a bologna sandwich, break it down into its ingredients:
        - Dempster's whole wheat bread- 2 slices
        - Bologna- 1 slice (50g)
        - Kraft mayonnaise (regular)- 1 tsp
        - Kraft cheese slice- 1 slice (30g)
    - For giving the size of meat like steak, use measurement: 2" long x 2" wide x 1" thick
    - For a bowl of cereal, don't forget the extras:
        - Kellogg's Rice Krispies- ½ cup (15 g)
        - 2% Milk- ¼ cup
        - Brown sugar- 2 level tsp.

- ✓ If you have any questions or concerns, please contact Colleen at (902) 442-2882 or (709) 468-7943 (collect) or via email at colleen.faulkner@msvu.ca.

Appendix D

IWK Debriefing Form

STUDY DEBRIEFING FORM

TITLE: The Relationship between Body Esteem and Adherence to Diet and Pancreatic Enzyme Replacement Therapy, in Adolescents with Cystic Fibrosis (CF).*

SHORT TITLE: The Link between Feelings about the Body and Eating Habits and Taking Enzymes, Vitamins and Nutritional Supplements, in Teenagers with Cystic Fibrosis.*

MAIN RESEARCHER: Colleen Faulkner
2060 Quingate Pl. Apt 1115
Halifax, NS B3L 4P7
(902)442-2882/(709)468-7943
colleen.faulkner@msvu.ca

ASSOCIATE RESEARCHER: Janette Taper
Room 311, Evaristus Hall
MSVU, Halifax, NS B3M 2J6
(902) 457-6256 (w)
janette.taper@msvu.ca

SPONSOR: Mount Saint Vincent University.

You are invited to join a research study. The study is being conducted by a student at Mount Saint Vincent University. This study is being done to find out better ways of caring for people with cystic fibrosis (CF).

It is important that you understand the purpose of this study. Please read this information carefully with your parent/guardian. You do not have to take part. Taking part is entirely voluntary (your choice). If you have any questions that this form does not answer, the researcher will be happy to give you further information.

1. Introduction/Background:

People with cystic fibrosis (CF) have special diet and health needs. It is normal for those with CF to find it hard to eat all that they are told to. It is also normal, from time to time, for those with CF to skip taking some of their drugs, for different reasons. As a teen, having CF may be hard. Sometimes there is pressure for teenagers to be very thin, like those seen in magazines and on television. While we all may want to look like this from time to time, trying to be thin may be dangerous for those with CF.

Can I be in the study?
You CAN take part in the study if you answer YES to all these questions:

- Are you between the ages of 13 and 20, inclusive?
- Do you have cystic fibrosis?
- Do you go to the clinic at the IWK?

- Do you take, or have you been asked to take, enzymes when you eat?

BUT, you CANNOT take part if you answer YES to any of these questions:

- Do you have diabetes?
- Has a doctor said that you have an eating disorder?
- Are you pregnant or breastfeeding/nursing?

How do I give permission to be in the study?

- When you mail back the completed forms you have given your permission to take part in the study.

2. Purpose of the study:

The purpose of the study is to find out how teens with CF feel about their bodies, to find out about their eating habits and find out if they miss doses of medication and/or nutritional supplements.

3. Study Design:

You have been identified as having cystic fibrosis by the nurse coordinator of your CF clinic. This is a multi-centered study to be carried out at the IWK in Halifax and the Janeway, in Newfoundland. Around 40-50 people will get a package. It is hoped that at least 10 people will participate.

Please read this "Debriefing Form" with a parent/guardian to help you decide if you want to take part in the study. Take as much time as you need to decide.

4. What participation involves:

If you decide to take part you will be asked to:

- Read and understand this "Debriefing Form";
- Write down the ID Code on your surveys, in case you decide to withdraw later on;
- Complete the following surveys:
    - "The Body-Esteem Scale For Adolescents and Adults";
    - "The Eating Attitudes Test";
    - "The Adherence Survey";
    - A "Three-Day Food Diary";
- Complete the Communication of Results (CR) Card;
- Return all completed surveys in the envelope provided; and
- Contact Colleen if you have ANY questions or if you decide to quit the study.

Do I have to take part?

- NO, the choice to take part in the study is up to you and your parent/guardian

- You are free to choose not to take part in the study
- If you do decide to take part and later you change your mind, you can still withdraw
- No matter what you decide, your decision will be supported. Your current medical care will NOT be changed. No one will be upset with you if you decide not to take part or change your mind.

How long will it take to be in the study?

You will be in the study for a minimum of three (3) days, when you are doing the "Three-Day Food Diary". Answering the surveys should take no more than a total of one (1) hour.

4. Possible risks and discomforts:

You may find some questions on the surveys may make you upset or sad. You may not like all the questions that you will be asked. You do not have to answer questions if they upset you or make you sad. There is the possibility that some unforeseen harm may occur, but this is not likely. You can phone Colleen Faulkner at any time if you have any questions or concerns.

5. Benefits:

It is not known if this study will help you in any way. However, it may help others with CF.

6. Alternatives:

There are no other alternatives. You can choose to take part or not.

7. Withdrawal from Participation:

If you choose to take part in the study and later you change your mind, you are free to stop taking part at any time, even if you have mailed all the forms. If you decide to stop, nothing bad will happen, simply follow these steps:

- Write down the ID Code from the surveys;
- Call, email, or mail Colleen Faulkner;
- Tell her that you are in the CF study and want to quit;
- You DO NOT need to give your name, just give your ID Code;
- When Colleen gets your ID Code, she will look for those survey forms and they will not be used in the study.

8. Confidentiality:

If the results of the study are published, the publication will not contain any information which would identify you. Study records will be stored in a locked area and will be kept for 5 years post publication as required by the IWK Research Ethics Board.

9. Cost of Participation:
There is no cost for being in the study. Everything you need is in this package, including postage for the return. If you must phone Colleen, you may call collect.

None of your rights are waived.

To thank you for participating in the study, you can win one of two Empire Theatres gift certificates. All you have to do is check off the correct box on the Communication of Results Card (CR) and return it with your surveys. Participants at both Centers' who complete the survey and fill out the card will be included in the draw. Good luck!

10. What About Questions or Problems?

The researcher for this study is Colleen Faulkner. If you have any questions or concerns you may contact her at (902) 442-2882 or (709) 468-7943 (collect) or via email at colleen.faulkner@msvu.ca. You may also contact her supervisor Janette Taper at (902) 457-6256.

You can contact the dietitian at the IWK CF clinic, Sheri Rose at (902) 470-7875, or you can speak to someone who is not directly involved in the study by contacting the Acting Chair of the Mount Saint Vincent University Research Ethics Board by phone at (902) 457-6350 or the IWK Research Services Office at (902)470-8765.

* Note: Debriefing form used original study title.

Appendix E

Janeway Debriefing Form

STUDY DEBRIEFING FORM

TITLE: The Relationship between Body Esteem and Adherence to Diet and Pancreatic Enzyme Replacement Therapy, in Adolescents with Cystic Fibrosis (CF).*

SHORT TITLE: The Link between Feelings about the Body and Eating Habits and Taking Enzymes, Vitamins and Nutritional Supplements, in Teenagers with Cystic Fibrosis.*

MAIN RESEARCHER: Colleen Faulkner
2060 Quingate Pl. Apt 1115
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(902)442-2882/(709)468-7943
colleen.faulkner@msvu.ca

ASSOCIATE RESEARCHER: Janette Taper
Room 311, Evaristus Hall
MSVU, Halifax, NS B3M 2J6
(902) 457-6256 (w)
janette.taper@msvu.ca

SPONSOR: Mount Saint Vincent University.

You are invited to join a research study. The study is being conducted by a student at Mount Saint Vincent University. This study is being done to find out better ways of caring for people with cystic fibrosis (CF).

It is important that you understand the purpose of this study. Please read this information carefully with your parent/guardian. You do not have to take part. Taking part is entirely voluntary (your choice). If you have any questions that this form does not answer, the researcher will be happy to give you further information.

1. Introduction/Background:

People with cystic fibrosis (CF) have special diet and health needs. It is normal for those with CF to find it hard to eat all that they are told to. It is also normal, from time to time, for those with CF to skip taking some of their drugs, for different reasons. As a teen, having CF may be hard. Sometimes there is pressure for teenagers to be very thin, like those seen in magazines and on television. While we all may want to look like this from time to time, trying to be thin may be dangerous for those with CF.

Can I be in the study?
You CAN take part in the study if you answer YES to all these questions:

- Are you between the ages of 13 and 20, inclusive?
- Do you have cystic fibrosis?
- Do you go to the clinic at the Janeway?

- Do you take, or have you been asked to take, enzymes when you eat?

BUT, you CANNOT take part if you answer YES to any of these questions:

- Do you have diabetes?
- Has a doctor said that you have an eating disorder?
- Are you pregnant or breastfeeding/nursing?

How do I give permission to be in the study?

- When you mail back the completed forms you have given your permission to take part in the study.

2. Purpose of the study:

The purpose of the study is to find out how teens with CF feel about their bodies, to find out about their eating habits and find out if they miss doses of medication and/or nutritional supplements.

3. Study Design:

You have been identified as having cystic fibrosis by the nurse coordinator of your CF clinic. This is a multi-centered study to be carried out at the IWK in Halifax and the Janeway. Approximately 40-50 people will get a package- it is hoped that at least 10 will take part.

Please read this "Debriefing Form" with a parent/guardian to help you decide if you want to take part in the study. Take as much time as you need to decide.

4. What participation involves:

If you decide to take part you will be asked to:

- Read and understand this "Debriefing Form";
- Write down the ID Code on your surveys, in case you decide to withdraw later on;
- Complete the following surveys:
    - "The Body-Esteem Scale For Adolescents and Adults";
    - "The Eating Attitudes Test";
    - "The Adherence Survey";
    - A "Three-Day Food Diary";
- Complete the Draw Card, if desired;
- Return all completed surveys in the envelope provided; and
- Contact Colleen if you have ANY questions or if you decide to quit the study.

Do I have to take part?

- NO, the choice to take part in the study is up to you and your parent/guardian
- You are free to choose not to take part in the study

- If you do decide to take part and later you change your mind, you can still withdraw
- No matter what you decide, your decision will be supported. Your current medical care will NOT be changed. No one will be upset with you if you decide not to take part or change your mind.

How long will it take to be in the study?

You will be in the study for a minimum of three (3) days, when you are doing the "Three-Day Food Diary". Answering the surveys should take no more than a total of one (1) hour.

4. Possible risks and discomforts:

You may find some questions on the surveys may make you upset or sad. You may not like all the questions that you will be asked. You do not have to answer questions if they upset you or make you sad. There is the possibility that some unforeseen harm may occur, but this is not likely. You can phone Colleen Faulkner at any time if you have any questions or concerns.

5. Benefits:

It is not known if this study will help you in any way. However, it may help others with CF.

6. Alternatives:

There are no other alternatives. You can choose to take part or not.

7. Withdrawal from Participation:

If you choose to take part in the study and later you change your mind, you are free to stop taking part at any time, even if you have mailed all the forms. If you decide to stop, nothing bad will happen, simply follow these steps:

- Write down the ID Code from the surveys;
- Call, email, or mail Colleen Faulkner;
- Tell her that you are in the CF study and want to quit;
- You DO NOT need to give your name, just give your ID Code;
- When Colleen gets your ID Code, she will look for those survey forms and they will not be used in the study.

8. Liability statement:

When the completed forms are received, you are saying that you understand the information about the research study and are willing to participate. When you mail back the completed forms, you do not give up your legal rights. Researchers or agencies involved in this research study still have their legal and professional responsibilities.

9. Confidentiality:

Your privacy is very important. The following things will be done to make sure no one can find out that you are taking part in the study:

- You DO NOT need to put your name, address, or phone number on ANY of the forms;
- ID Codes are used on the surveys that you will return. This way, we do not know who filled out the forms but we can keep all your forms together;
- There will be no way to identify participants when results are written up and if they are published. No ID Codes will be referred to;
- The person who mailed you this package does know who you are, however, s/he will not know if you decide to take part and will not see any returned forms; and
- All forms and data will be stored in a locked cabinet or on a password protected computer.

10. Questions:

You can ask Colleen Faulkner for more information on the study or if you have any questions or concerns. She can be reached by mail, email, or by calling (902) 442-2882 or (709) 468-7943 collect, or by emailing colleen.faulkner@msvu.ca. You do not have to give your name-just tell her that you have a question about the CF study. You can also call the associate investigator, Janette Taper, at (902) 457-6256. You do not have to give your name-just tell Colleen or Janette that you have a question about the CF study.

You can also talk to someone who is not involved with the study at all, but can let you know your rights as a participant in a research study. This person can be reached through: Office of the Human Investigation Committee (HIC) at (709) 777-6974 or by Email at hic@mun.ca. You can also contact the Acting Chair of the Mount Saint Vincent University Research Ethics Board by phone at (902) 457-6350.

To thank you for participating in the study, you can win one of two Empire Theatres gift certificates. All you have to do is check off the correct box on the Communication of Results Card (CR) and return it with your surveys. Participants at both Centers' who complete the survey and fill out the card will be included in the draw. Good luck!

* Note: Debriefing form used original study title.

Appendix F

Three-Day Food Diary

Time of Day	Foods Eaten	Amount Eaten	Specific Preparation (if any)	Supplements (if any) (Include brand & amount here)	# Enzymes Taken (if any)
Breakfast					
Morning Snack					
Lunch					

Time of Day	Foods Eaten	Amount Eaten	Specific Preparation (if any)	Supplements (if any) (Include brand & amount here)	# Enzymes Taken (if any)
Mid-Afternoon Snack					
Supper					
Evening Snack					

Appendix G

The Eating Attitudes Test-26

Please think about how often you agree with the following statements, ranging from “always” to “never”. Place a check mark in the box beside your response.

	Always	Usually	Often	Sometimes	Rarely	Never
1. Am terrified about being overweight						
2. Avoid eating when I am hungry						
3. Think about food often						
4. Have gone on eating binges where I feel I may not be able to stop						
5. Cut my food into small pieces						
6. Aware of the calorie content of foods I eat						
7. Avoid foods that are high in carbohydrates (bread, rice, potatoes, etc.)						
8. Feel that others would like it if I ate more						
9. Throw-up after I have eaten						
10. Feel extremely guilty after eating						
11. Think a lot about wanting to be thinner						
12. Think about burning up calories when I exercise						
13. Other people think I'm too thin						
14. Think about having fat on my body a lot						
15. Take longer than others to eat my meals						
16. Avoid foods with sugar in them						
17. Eat diet foods						
18. Feel that food controls my life						

	Always	Usually	Often	Sometimes	Rarely	Never
19. Can control myself well around food						
20. Feel that others pressure me to eat						
21. Give too much time and thought to food						
22. Feel uncomfortable after eating sweets						
23. Take part in dieting behaviors						
24. Like my stomach to be empty						
25. Have the urge to throw-up after meals						
26. Enjoy trying new rich foods (foods that are full, heavy, creamy, etc)						

Appendix H

The Body Esteem Scale for Adolescents and Adults

Please think about how often you agree with the following statements, ranging from "never" to "always". Place a check mark in the box beside your response.

	Never	Seldom	Sometimes	Often	Always
1. I like what I look like in pictures					
2. Other people consider me good looking					
3. I am proud of my body					
4. I am always thinking about trying to change my body weight					
5. I think my appearance would help me get a job					
6. I like what I see when I look in the mirror					
7. There are lots of things I'd change about my looks if I could					
8. I am happy with my weight					
9. I wish I looked better					
10. I really like what I weigh					
11. I wish I looked like someone else					
12. People my own age like my looks					
13. My looks upset me					
14. I'm as nice looking as most people					
15. I'm pretty happy about the way I look					
16. I feel I weigh the right amount for my height					
17. I feel ashamed of how I look					
18.Weighing myself depresses me					
19. My weight makes me unhappy					
20. My looks help me to get dates					
21. I worry about the way I look					
22. I think I have a good body					
23. I'm looking as nice as I'd like to					

Appendix I

The Adherence Survey

People with CF have special food needs. They are often asked to eat a lot of food and this may be hard to do. If those with CF can't eat all they need to, a doctor or dietitian may ask them to take a supplement. People with CF usually take a lot of pills each day and may miss some. I would like to know how often this happens to you and if there are any problems that stop you from taking your pills or nutritional supplements.

Section 1: Vitamins

Many of those with CF are asked to take vitamins by their doctor or dietitian. In this section, I would like to know if you are currently taking vitamins, what vitamin(s) you are taking, and if there are any problems that stop you from taking them.

NOTE: The term *Full Dose* means the total amount of vitamins that your doctor or dietitian has asked you to take EACH day.

1. Has your doctor or dietitian asked you to take vitamins?
   - o Yes
   - o No (If NO, please go to section 2)

2. Do you currently take the vitamins your doctor or dietitian asked you to take?
   - o Yes, I currently take ALL of the vitamins my doctor/dietitian asked me to take
   - o Yes, I currently take SOME of the vitamins my doctor/dietitian asked me to take
   - o I DO NOT currently take any vitamins my doctor/dietitian asked me to take (If NO, please go to question 7)

3. Please list each KIND of vitamin that you currently take and the amount your doctor or dietitian asked you to take in the chart below.

   Example: Leisha was asked to take:
   2 Vitamin A pills,
   1 Vitamin E pill, and
   2 ADEKS® pills each day.

   The *Full Dose* of vitamins she was asked to take is 5 pills each day.

Kind of Vitamin	Amount Asked to Take

Your *Full Dose* of vitamins= _____
(add the total number of vitamins together)

4. How many days a week do you usually take ALL of the Full Dose of vitamins that your doctor or dietitian asked you to take?

   Example: The *Full Dose* of vitamins Leisha was asked to take is 5 pills each day. She usually takes the *Full Dose* of 5 pills on 3-4 days a week.

   - o I never take a *Full Dose* of vitamins
   - o I take a *Full Dose* of vitamins 1-2 days a week
   - o I take a *Full Dose* of vitamins 3-4 days a week
   - o I take a *Full Dose* of vitamins 5-6 days a week
   - o I take a *Full Dose* of vitamins every day of the week

5. How many days a week do you usually take HALF OR MORE of the Full Dose of vitamins your doctor or dietitian asked you to take?

   Example: The *Full Dose* of vitamins Leisha was asked to take is 5 pills each day. HALF of the *Full Dose* is 2 and a ½ pills, or 2 pills (round down to the nearest whole number). She usually takes 2 pills OR MORE, on 5-6 days a week.

   - o I never take HALF OR MORE of the *Full Dose* of vitamins
   - o I take HALF OR MORE of the *Full Dose* of vitamins 1-2 days a week
   - o I take HALF OR MORE of the *Full Dose* of vitamins 3-4 days a week
   - o I take HALF OR MORE of the *Full Dose* of vitamins 5-6 days a week
   - o I take HALF OR MORE of the *Full Dose* of vitamins every day of the week

6. Please check ALL the reasons why you miss any vitamins you were asked to take by your doctor or dietitian, or why you do not take them at all.

   - o Sometimes I tend to forget to take them
   - o I do not know why I have to take vitamins
   - o I do not like to take them in front of others
   - o They taste awful
   - o I have problems swallowing them
   - o Taking the vitamins does not make me feel better
   - o Taking the vitamins makes me feel worse
   - o It takes a lot of effort or time to take them
   - o I cannot afford to buy them
   - o Other _____

Section 2: Nutritional Supplements

Some people with CF have been asked to take a nutritional supplement by their doctor or dietitian. These supplements may help them gain weight. In this section, I would like to know about any nutritional supplements that you take, how many you take, and if there are any problems that stop you from taking them.

Nutritional supplements include:

- Shakes such as Boost®, Ensure®, Pediasure®, and Scandishake®.
- Other products such as special puddings or bars
- Tube feedings such as NG-Tubes, G-Tubes, or J-Tubes

NOTE: If you are using tube feedings, try to give the name of the formula you use and how often you use it. If you do not know the name of the formula, simply write how often you use tube feedings each day.

NOTE: The term *Full Dose* means the total amount of nutritional supplements that your doctor or dietitian has asked you to take EACH day.

7. Has your doctor or dietitian asked you to take any nutritional supplements?
   - Yes
   - No (If NO, go to section 3)

8. Do you currently take the nutritional supplement(s)?

   - Yes, I currently take ALL of the nutritional supplements my doctor/dietitian asked me to take
   - Yes, I currently take some of the nutritional supplements my doctor/dietitian asked me to take
   - I do not currently take any of the nutritional supplements my doctor/dietitian asked me to take (If checked please go to question 14)

9. Please list each KIND of nutritional supplement that you currently take and the amount your doctor or dietitian asked you to take in the chart below. Adding the total number together will give you the *full dose* of supplements you were asked to take each day.

   Example: Tim was asked to take:
   2 cans of Boost® a day and
   1 cup of Scandishake® a day.

   The *Full Dose* of nutritional supplements he was asked to take is 3 supplements each day.

Kind of Nutritional Supplement	Amount Asked to Take

Your *Full Dose* of nutritional supplements= _____
(Add the total number of supplements together)

10. How many days a week do you usually take ALL of the *Full Dose* of nutritional supplements that your doctor or dietitian asked you to take?

    Example: The *Full Dose* of nutritional supplements Tim was asked to take is 3 supplements each day. He usually takes the *Full Dose* of 3 supplements on 5-6 days a week.

    - o I never take a *Full Dose* of nutritional supplements
    - o I take a *Full Dose* of nutritional supplements 1-2 days a week
    - o I take a *Full Dose* of nutritional supplements 3-4 days a week
    - o I take a *Full Dose* of nutritional supplements 5-6 days a week
    - o I take a *Full Dose* of nutritional supplements every day of the week

11. How many days a week do you usually take HALF OR MORE of the *Full Dose* of nutritional supplements your doctor or dietitian asked you to take?

    Example: The *Full Dose* of nutritional supplements Tim was asked to take is 3 each day. HALF of the *Full Dose* is 1 and a ½ supplements, or 1 supplement (round down to the nearest whole number). He usually takes 1 OR MORE nutritional supplements, on every day of the week.

    - o I never take HALF OR MORE of the *Full Dose* of nutritional supplements
    - o I take HALF OR MORE of the *Full Dose* of nutritional supplements 1-2 days a week
    - o I take HALF OR MORE of the *Full Dose* of nutritional supplements 3-4 days a week
    - o I take HALF OR MORE of the *Full Dose* of nutritional supplements 5-6 days a week
    - o I take HALF OR MORE of the *Full Dose* of nutritional supplements every day of the week

12. Please check ALL the reasons why you miss any supplements that you were asked to take by your doctor or dietitian, or why you do not take them at all.

    - o Sometimes I tend to forget to take them
    - o I do not know why I need to take nutritional supplements
    - o I do not like to take nutritional supplements in front of others
    - o The supplement(s) tastes awful
    - o I feel that taking the supplement(s) does not make my health any better
    - o Taking the supplement(s) makes me feel worse
    - o It takes a lot of effort or time to take the nutritional supplement(s)
    - o I cannot afford to get the supplement(s)
    - o I am allergic to the supplement(s)
    - o I am happy at my current weight
    - o Other_____

Section Three: Pancreatic Enzyme Replacement Therapy

Most people with CF have to take enzymes when they eat to digest their food. In this section, I would like to know if you take any enzymes, how often you take them, and if you have any problems which stop you from taking them.

NOTE: The term *Full Dose* means the amount of enzymes that your doctor or dietitian has asked you to take with a meal and snack.

13. How many enzymes did your doctor and/or dietitian and/or nurse ask you to take with:

    - A Meal? _____
      (This is your *Full Dose* of enzymes with a meal)

    - A Snack? _____
      (This is your *Full Dose* of enzymes with a snack)

14. How many days a week do you usually take ALL of the *Full Dose* of enzymes that your doctor/dietitian/nurse asked you to take WITH A MEAL?

    Example: The *Full Dose* of enzymes Bette was asked to take with a meal is 6 pills. She usually takes the *Full Dose* of 6 pills with a meal on 1-2 days a week.

    - I never take a *Full Dose* of enzymes with a meal
    - I take a *Full Dose* of enzymes with a meal 1-2 days a week
    - I take a *Full Dose* of enzymes with a meal 3-4 days a week
    - I take a *Full Dose* of enzymes with a meal 5-6 days a week
    - I take a *Full Dose* of enzymes with a meal every day of the week

15. How many days a week do you usually take HALF OR MORE of the *Full Dose* of enzymes your doctor/dietitian/nurse asked you to take WITH A MEAL?

    Example: The *Full Dose* of enzymes Bette was asked to take with a meal is 6 pills. HALF of the *Full Dose* is 3 pills. She usually takes 3 OR MORE pills with a meal, on 3-4 days of the week.

    - I never take HALF OR MORE of the *Full Dose* of enzymes with a meal
    - I take HALF OR MORE of the *Full Dose* of enzymes with a meal 1-2 days a week
    - I take HALF OR MORE of the *Full Dose* of enzymes with a meal 3-4 days a week
    - I take HALF OR MORE of the *Full Dose* of enzymes with a meal 5-6 days a week
    - I take HALF OR MORE of the *Full Dose* of enzymes with a meal every day of the week

16. How many days a week do you usually take ALL of the *Full Dose* of enzymes that your doctor/dietitian/nurse asked you to take WITH A SNACK?

    Example: The *Full Dose* of enzymes Bette was asked to take with a snack is 3 pills. She usually takes the *Full Dose* of 3 pills with a snack, on 5-6 days a week.

    - o I never take a *Full Dose* of enzymes with a snack
    - o I take a *Full Dose* of enzymes with a snack 1-2 days a week
    - o I take a *Full Dose* of enzymes with a snack 3-4 days a week
    - o I take a *Full Dose* of enzymes with a snack 5-6 days a week
    - o I take a *Full Dose* of enzymes with a snack every day of the week

17. How many days a week do you usually take HALF OR MORE of the *Full Dose* of enzymes your doctor/dietitian/nurse asked you to take WITH A SNACK?

    Example: The *Full Dose* of enzymes Bette was asked to take with a snack is 3 pills. HALF of the *Full Dose* is 1 and a ½, or 1 pill (round down to the nearest whole number). She usually takes 1 OR MORE pills with a snack, on every day of the week.

    - o I never take HALF OR MORE of the *Full Dose* of enzymes with a snack
    - o I take HALF OR MORE of the *Full Dose* of enzymes with a snack 1-2 days a week
    - o I take HALF OR MORE of the *Full Dose* of enzymes with a snack 3-4 days a week
    - o I take HALF OR MORE of the *Full Dose* of enzymes with a snack 5-6 days a week
    - o I take HALF OR MORE of the *Full Dose* of enzymes with a snack every day of the week

18. Please check ALL the reasons why you miss any enzymes that you were asked to take by your doctor/dietitian/nurse with meals AND snacks, or why you do not take them at all.

    - o I tend to forget to take them at times
    - o I do not know why I need to take enzymes
    - o I do not like to take them in front of others
    - o I have trouble swallowing them
    - o I feel that taking enzymes does not make my health any better
    - o Taking enzymes makes me feel worse
    - o It takes a lot of effort or time to take them
    - o I cannot afford to get them
    - o I am happy at my current weight and do not want to gain weight
    - o I get side effects from the enzymes
    - o I adjust the amount of enzymes I take depending on what I eat
    - o Other_____

Section 4: Demographic Information

The following section is for statistical purposes only. You DO NOT need to write your name on ANY of the forms of this questionnaire. If you do not want to give this information you do not have to.

19. How old are you? _____

20. Are you male or female? Male____ Female____

21. How much do you weigh? _____

22. How tall are you? _____

Appendix J

The Communications of Results Card

&

The Draw Card

COMMUNICATION OF RESULTS CARD (CR) (IWK)

Research results will be made available to you at the completion of the study. Would you like to receive a copy of the results?

- o Yes
- o No

Would you like to be included in the draw for one of two Empire Theatres® gift certificates?

- o Yes
- o No

Address (if you answered yes to any of the above):

THE DRAW CARD (JANEWAY)

If you would like to be included in a draw for one of two Empire Theatre® Gift Certificates, write your address on the card and include it in the envelope when you mail your surveys. Good-Luck! ~Colleen Faulkner

INCLUDE ME IN THE DRAW:

- o Yes
- o No

ADDRESS:

Appendix K

IWK Reminder Letter

Dear Participant,

Last month a package containing surveys seeking your opinion about CF, eating habits, taking your medication, and how you feel about your body, was mailed to you. Your name was selected since you attend the CF clinic at the IWK Health Centre.

If you have already completed and returned it to me, please accept my sincere thanks. If not, do so today. Because it has been sent out to only a small group of people with CF, it is **extremely** important that yours be included in the study if the results are to accurately represent the opinions of individuals with CF in the Atlantic region of Canada.

If, by some chance, you did not receive the package or have misplaced it and would like another, please call me right away, collect (902) 442-2882, and I will get another one in the mail to you today.

Don't forget that your participation will put you in a draw for 1 or 2 Empire Theatres ® gift certificates. If you have ANY questions about the study or the surveys, you can contact me, Colleen, or Janette Taper, who is my thesis advisor, or Sheri Rose, the dietitian at your clinic.

Sheri can be reached at:
- ✓ IWK Health Centre
  5850/5980 University Ave
  P.O. Box 3070
  Halifax, NS B3J 3G9
  (902) 470-7875

Colleen can be reached at:
- ✓ 2060 Quingate Place
  Apt 1115
  Halifax, NS B3L 4P7
  (902) 442-2882/ (709) 468-7943
  62Hcolleen.faulkner@msvu.ca

Janette can be reached at:
- ✓ Room 311, Evaristus Hall
  MSVU, Halifax, NS
  B3M 2J6
  (902) 457-6256 (w)
  61Hjanette.taper@msvu.ca

Sincerely,

Colleen Faulkner

Sheri Rose, B.Sc., P.Dt.

Appendix L

Janeway Reminder Letter

Dear Participant,

Last month a package containing surveys seeking your opinion about CF, eating habits, taking your medication, and how you feel about your body, was mailed to you. Your name was selected since you attend the CF clinic at the Janeway.

If you have already completed and returned it to me, please accept my sincere thanks. If not, do so today. Because it has been sent out to only a small group of people with CF, it is **extremely** important that yours be included in the study if the results are to accurately represent the opinions of individuals with CF in the Atlantic region of Canada.

If, by some chance, you did not receive the package or have misplaced it and would like another, please call me right away, collect (902) 442-2882, and I will get another one in the mail to you today.

Don't forget that your participation will put you in a draw for 1 or 2 Empire Theatres ® gift certificates. If you have ANY questions about the study or the surveys, you can contact me, Colleen, or Janette Taper, who is my thesis advisor, or Betty Sheppard, the nurse coordinator at your clinic.

Betty can be reached at:
✓ Betty Sheppard
Room 2J732, HSC
300 Prince Phillip Dr.
St. John's, NL A1B 3V6
(709) 777-4389
67HBetty.Sheppard@hccsj.nl.

Colleen can be reached at:
✓ 2060 Quingate Place
Apt 1115
Halifax, NS B3L 4P7
(902) 442-2882/ (709) 468-7943
68Hcolleen.faulkner@msvu.ca

Janette can be reached at:
✓ Room 311, Evaristus Hall
MSVU, Halifax, NS
B3M 2J6
(902) 457-6256 (w)
69Hjanette.taper@msvu.ca

Sincerely,

Colleen Faulkner

Betty Sheppard, R.N.

Appendix M

Nutrient Intakes of Participants

Table 11: Nutrient Intakes of Participants

	Caloric[a] Intake (Cal/ day)	Fat Intake[a] (g/day)	Protein Intake[a] (g/day)	Carbohydrate Intake[a] (g/day)	Percent Calories from Fat	Percent Calories from Protein	Percent Calories from Carbohydrates	EER[b] (Cal/ day)	EER + 20%[c] (Cal/ day)
Participant[d]									
A	2629	79	102	336	37.48%	20.75%	39.08%	2444	3472
B	2439	86	94	338	34.03%	12.52%	55.80%	NA[e]	NA
C	1042	43	54	102	37.11%	18.81%	41.48%	2862	4054
D	2997	139	102	102	36.70%	20.07%	43.24%	2795	3955
E	2426	92	76	338	29.82%	14.72%	55.95%	1436	2051
F	2011	83	95	209	27.20%	15.45%	51.17%	2206	3108
G	1401	57	70	151	41.65%	13.63%	44.15%	2701	3812
H	1875	62	69	262	31.61%	15.36%	55.43%	1999	2818
I	2748	109	92	354	35.76%	13.45%	51.50%	2320	3274

a. Intake is the average of total intakes for three days from Three-Day Food Diary. All calorie values are rounded to the nearest number.
b. EER= Estimated Energy Requirement with a physical activity level of Low Active (PAC of 1.13 for males and 1.16 for females).
c. EER+20%: In theory, the lowest recommended intake for those with CF, assuming a physical activity level of Low Active (20% added onto base estimated requirement).
d. Participants listed randomly and assigned a letter name of no value or significance.
e. Inadequate data provided to determine