Physiological and Perceptual Responses during Self-Regulated Exercise in Children with Cystic Fibrosis

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MSc

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Declaration

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Abstract

Purpose: To examine the physiological and perceptual responses during self-regulated exercise in children with cystic fibrosis (CF).

Methods: Twelve children (10.8 ± 2.0 y) with CF made 3 separate visits to the research laboratory in the school of Health and Human Performance in DCU. The first visit was used to determine VO_2peak and to anchor the children's OMNI RPE scale. The second visit was used to familiarise the children with the testing procedures. During the final visit children exercised for 20 min on a cycle ergometer. The exercise intensity was self-regulated. Heart rate (HR) and VO_2 were measured using telemetry and open circuit spirometry, respectively. Undifferentiated RPE (RPE-O) and differentiated RPE for the chest (RPC-C) and legs (RPE-L) were assessed using the Children's OMNI RPE scale.

Results: Perceptual and physiological responses remained stable after the first 5 min of exercise. Subjects exercised at 53% VO_2peak and 73% HRpeak during the final 15 min of self regulated exercise. RPE-O was 5.8 ± 2.6, RPE-C was 4.5 ± 2.6, and RPE L was 5.7 ± 2.4. The %VO_2peak and %HRpeak corresponding to the ventilatory breakpoint (Vpt) was 63% and 82% respectively. Two thirds of the subjects selected an exercise intensity ±20% VO_2@Vpt, and one third selected an intensity 31 - 38% below VO_2@Vpt.

Conclusion: Although the mean %VO_2peak was within the moderate to vigorous intensity range recommended for health and fitness benefits, there were large intra individual variations in RPE, %VO_2peak, %HRpeak and %Vpt during self regulated exercise in children with CF.

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CHAPTER I

INTRODUCTION

Cystic fibrosis (CF) is the most common, inherited, and life-shortening disease among the Caucasian population. It is caused by a mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene. The genetic defect leads to bronchiolitis, bronchitis, bronchiectasis, and eventually to fibrosis and irreversible loss of pulmonary function (12, 25), which limits exercise capacity and quality of life.

Aerobic capacity is a strong predictor of survival and quality of life in CF patients (85, 61, 49). Nixon et al (61) found that the CF patients with the highest aerobic fitness levels had a survival rate of 83% at 8 years of age compared to 51% at the same age in patients with the lowest fitness levels. Promoting regular physical activity to maintain or improve aerobic fitness could potentially increase the quality and length of life of CF patients (53). Surprisingly, parents and teachers are often concerned about children with CF participating in vigorous activities. Boas et al (60) found that parents of children with CF perceived exercise in a less positive light than parents with healthy children. These concerns may be transmitted to the children who in turn begin to question their own ability to participate in regular moderate-vigorous physical activities.

Participation in regular physical activity is influenced by a number of factors. These include demographic, psychological, behavioural, sociocultural, environmental and activity characteristics (64). Regular physical activity is strongly associated with enjoyment of exercise and participation in low-to-moderate levels of activity (64).

An exercise prescription can be defined as a specific guide provided to an individual in order to achieve his or her own measure of success or improvement. The exercise dose is usually characterized by the intensity, frequency, duration, progression and mode of exercise. The intensity of exercise is usually based on physiological (heart rate, oxygen uptake, blood lactate) responses during a submaximal, or maximal exercise test. However, exercise prescription based on meeting a target physiological response may be perceived as highly controlling and may exceed an individual's preferred level of intensity resulting in the establishment of negative attitudes toward physical activity (21). In contrast, allowing individuals to use effort perception to self-regulate exercise intensity may increase enjoyment and participation levels (93). This is not surprising considering that exertional feedback is often used unconsciously to regulate the pace of many daily activities. In a recent study Johnson et al (2006) found that 86% of women involved in aerobic exercise used effort perception exclusively to determine exercise intensity (34).

The perception of exertion is defined as the intensity of effort, strain, discomfort or fatigue that a person experiences during exercise (70). An individual's rating of perceived exertion (RPE) integrates the responses from the peripheral working muscles and joints, the central cardiovascular system, the respiratory system and the central nervous system (7).

A number of scales have been developed to assess RPE (7). The most widely used is the Borg 15-category scale. This scale was developed for use in adults, and is comprised of a numerical scale that ranges from 6 – 20 and verbal descriptors that represent the intensity of physical exertion**.** The Borg Scale can present methodological and semantic limitations when used by children and adolescents. Some children, particularly those under the age of 11 are unable to consistently assign words to numbers or phrases that represent exercise related feelings. In addition, they may find it difficult to use the verbal cues that are not part of their current vocabulary. The OMNI scale was recently developed to overcome these limitations. The OMNI scale uses a series of pictures depicting individuals exercising at increasing exercise intensities. The pictures are arranged along a numerical scale from 0 to 10 and verbal cues are placed at equal intervals along the scale (71).

The majority of studies that have evaluated self-regulated exercise have used walking (57, 67, 87, 16). In general, these studies found that men and women selected exercise intensities that ranged from 52 - 71% VO_2max, 67 – 81% HRmax and an RPE of "fairly light" (10.9 - 11.6) on the Borg 15-category RPE scale.

When using self-regulated exercise intensity, it is important to ensure that the chosen intensity elicits a physiological response likely to improve cardiovascular health. The purpose of this study was to compare physiological and perceptual responses during self-regulated exercise in children with CF. It is hypothesized that children with CF will self-select an intensity that is; i) within the stimulus response range to maintain or improve cardiovascular fitness, ii) at or slightly below the ventilatory breakpoint (Vpt) and iii) corresponds to an RPE between 4 – 6 on the OMNI RPE scale

CHAPTER II

LITERATURE REVIEW

Introduction

Cystic fibrosis (CF) is an autosomal recessive genetic disease that occurs in one of every 3,200 Caucasian births. Males and females are equally affected. However, Ireland with a rate of one in 1600 births has the highest incidence of CF worldwide. There are 1,143 CF patients in Ireland of which 45% are adults and 55% children (91). The development of sensitive diagnostic techniques and better supportive treatment in multidisciplinary CF care centres, have dramatically increased life expectancy of cystic fibrosis patients. The life expectancy CF patients has increased from 10 years in the 1960's to 37.4 years in 2007 (11).

CF is caused by mutations in the cystic fibrosis transmembrane regulator (CFTR) gene on the long arm of chromosome 7 (70). The CFTR gene codes for a protein that spans the apical membrane of epithelial cells lining the airways, pancreatic ducts, sweat ducts, intestines, biliary tree, and vas deferens, and normally regulates the transport of chlorine and other ions. The chloride ion channel is important in producing sweat, mucus and digestive juices.

Normal levels of mucus provide the lungs with a protective mechanism to reduce the risk of infections. The mucus traps bacteria, viruses, fungi, and other agents that may cause disease. These particles pass back out of the lungs into the throat where they can be coughed up or swallowed and digested. In individuals with CF, the absence of this chloride ion channel affects the ability to transport salt and water into and out of cells, which in turn results in the production of dry, thick and sticky secretions that impair mucociliary clearance (90). A combination of mucus plaques and impaired mucociliary clearance establishes a breeding ground for bacterial infection (90).

Pseudomonas aeruginosa, staphylococcus aureus, haemophilus influenzae and stenotrophomonas maltophilia (70) are the predominant pathogens causing chronic lung infections in patients with CF. The resulting inflammation and swelling along with excessive mucus accumulation may decrease the diameter of the airways, and make it difficult to move air in and out of the lungs. Although antibiotics may help to contain infections within the lungs, they usually fail to eradicate the organisms, and destruction of the airways is continual, eventually leading to bronchiolitis, bronchitis, bronchiectasis and eventually fibrosis and irreversible loss of pulmonary function (60). Chronic lung infection is the principle cause of death in >90% of CF patients. CF also increases susceptibility to malnutrition, pancreatic, liver and gall bladder disease, diabetes, male infertility, and poor growth (70). Symptoms vary depending on age, prior treatment, types of infections encountered, environmental factors and the extent to which various organs are affected (70).

The progression of pulmonary disease plays an important role in determining the clinical outcome in CF patients. Pulmonary function declines by 2-3 % per year in adult CF patients (66). A decline is also found in children between the ages of 6-18 years (29). Forced expiratory volume in 1 second (FEV1), the volume of air that can be forcibly expired in one second following a deep inspiration, is a good marker of pulmonary disease severity (40). The FEV1 value is expressed as a percentage of a predicted value and is adjusted for age, height, and gender. CF is classified as mild when the FEV1 is between 60-80% of predicted, moderate between 30-60% of predicted and severe <30% of predicted.

Treatments for CF include a wide range of therapies based on current infection(s) and disease progression, and include drug based therapies, airway clearance mechanisms, implanted devices and nutritional interventions. The severity of the illness increases with age, and treatments become progressively more expensive and more difficult to manage. Patients with more severe CF symptoms may require hospitalization for organ transplantation (43) and palliative care. Specialist CF treatment is currently provided in a number of centre's throughout Ireland (91).

Exercise Limitations and CF

Exercise intolerance is a characteristic of CF. A number of factors including pulmonary complications, impaired nutrition, metabolic abnormalities, limb muscle dysfunction (48, 41) and psychosocial factors (28), contribute to an increased intolerance to exercise in CF patients, and often negates exercise participation (97). Adults with CF have lower levels of physical activity than their healthy age matched counterparts due in part to the fact that the number of complications increases with age.

Pulmonary function inefficiency may be caused by abnormal pulmonary mechanics and problems with gaseous exchange in the lungs (52) resulting in increased minute ventilation during exercise, and greater sensations of dyspnoea. Airway remodeling that occurs in CF, increases airway resistance, ventilation/perfusion mismatching and the work of breathing. Respiratory muscles are repeatedly exposed to periods of increased work because of relative hyperinflation and alterations in the length tension relation during periods of hypoxia.

In apparently healthy individuals, ventilation rates at peak exercise are usually <60-70% of maximal voluntary ventilation (MVV). Among individuals with CF it is not uncommon for ventilation rates to reach MVV during maximal exercise while tidal volume remains low, indicating a mechanical ventilatory limitation to exercise. Although high percentages of MVV may mechanically limit performance, peripheral factors, chest pain or sensations of dyspnoea may limit peak exercise performance at lower percentages of MVV (59). A high respiratory rate coupled with a low tidal volume results in a high percentage of atmospheric air remaining in the airway dead space zone, and not participating in gaseous exchange. It is likely that CF patients are unable to compensate for the additional dead space due to the lack of sufficient ventilatory reserve. Inadequate uptake of oxygen in the lungs will limit aerobic exercise performance. Decreasing the intensity but increasing the duration of exercise reduces feelings of breathlessness and decreases the muscular effort of breathing to less than a third of the effort needed for higher intensity training (97).

Arterial hypoxemia caused by oxygen desaturation (52, 59) may also contribute to the decrease in exercise tolerance in CF patients. Oxygen saturation is >95% during exercise in healthy individuals, and may decrease to <90% in CF patients with an FEV1 ≤ 50% of predicted values (59). McKone et al (51) found that arterial hypoxemia limits maximal exercise in adult men and women with mild to severe CF.

When the oxygen supply is unable to keep up with the demand, there is an increased reliance on anaerobic metabolism with a concomitant increase in the production of lactic acid. Since lactic acid has a relatively low pKa (3.9), it is almost totally dissociated at the pH of muscle cells (~7.0). The increase in hydrogen ions (H^+) decreases intracellular pH, which has a deleterious effect on muscle function resulting in fatigue. The exercise intensity at which lactate production begins to exceed the rate of removal is termed the lactate threshold (LT).

Buffering of H^+ by intracellular bicarbonate increases the non-metabolic production of CO_2 (lactate + Na-bicarbonate→ Na-lactate, + H_2O + CO_2). Subsequent activation of peripheral chemoreceptors by increased levels of circulating CO_2, signals the inspiratory centre in the brain to increase ventilation (Ve) causing breathlessness (97). The exercise intensity at which Ve and VCO_2 increase more rapidly than O_2 is called the ventilatory breakpoint (Vpt).

Malnutrition, low body weight (caused by pancreatic insufficiency), a reduced appetite and vomiting caused by lung infections (70, 29) are other common problems that may limit exercise capacity in individuals with CF. Malnutrition and low body weight symptoms may be exacerbated by the fact that some CF patients have an elevated resting energy expenditure (54, 21), and a concomitant increase in the energy cost of exercise. The elevated resting energy expenditure may be caused by excessive airway inflammation and pulmonary exacerbations that increase the energy cost of breathing.

A defect in fat metabolism may also negatively impact on moderate intensity exercise in CF patients (35). Compared to healthy individuals, CF patients appear to have difficulties utilising fat stores during moderate intensity exercise, and rely to a greater degree on carbohydrate fuel sources for energy. There is speculation that the defects in fat metabolism during exercise are caused by the inefficient oxidation of fat in the mitochondria (35).

Weight loss and metabolic pancreatic problems associated with a decreased ability to exercise may be counteracted by an early diagnosis of the disease. Farrell et al (23) found that weight gain and early growth are better in patients diagnosed by neonatal screening compared to those who are diagnosed at a later time. Hyper alimentation that requires a daily calorie intake of 3,500 kcal results in an increase in BMI (31), which would subsequently help to improve work capacity and overall well-being.

Alterations in muscle function may affect exercise capacity and performance in CF patients. It is not clear whether the alterations in muscle function in CF patients are caused by intrinsic abnormalities in the skeletal muscles, or if they are due to inadequate nutritional status and metabolism problems. Some studies have reported normal muscle strength in CF patients, but decreased muscle mass due to inadequate nutrition (97). This view is corroborated by a study showing that weight training can increase both muscle strength and size (88) indicating that muscle weakness is due to inadequate nutrition and/or inadequate training and not an intrinsic inability to develop muscle force. However, other studies report that muscular weakness and decreased muscle mass in CF patients is indicative of intrinsic abnormalities in the skeletal muscles as opposed to the effects of diminished nutritional status or impaired metabolism (14).

Corticosteroids are part of the pharmacologic armamentarium commonly used to treat infections in CF patients. Pharmacological administration of corticosteroids causes nitrogen wasting and net muscle protein catabolism. This is in part mediated by the induction of the enzyme glutamine synthetase. Gene-mediated effects of corticosteroid treatments are evident in muscle after short-term administration (50).

Exercise Response and CF

Studies examining the physiological responses to exercise in CF patients began in the 1970's (27). Research in the late 1980's indicated that ventilation and mucus clearance could be improved with regular exercise (36, 101). Airway clearance techniques promote loosening and removal of excess mucus from the lungs in order to prevent infection and to improve lung function. Postural drainage and percussion requires a helper or therapist to tap the patient on the back and chest to free the mucus and remove it from the lungs. Exercise can provide a natural form of chest physiotherapy (97). The vibrations generated during exercise help to mobilise and expel mucus and accompanying pathogens from the lungs. Indeed exercise in conjunction with chest physiotherapy has been shown to be better than chest physiotherapy alone for improving pulmonary function and mucus clearance (9).

Physical activity levels among children appear to be related to the severity of CF. Nixon et al (60) found that prepubescent children with mild CF are more active than their healthy peers. In contrast, those with moderate to severe CF are less active than healthy children. The CF children in Nixon's study participated in 2.0 ± 2.5 h/wk of vigorous exercise compared with 3.7 ± 2.8 h/wk for healthy age matched controls. Children with CF are generally smaller with a lower body weight and less fat free mass than healthy children. Pubertal development is also delayed by ~2yrs in children with CF. For these reasons care should be taken when comparing the fitness levels of children with CF and their age matched healthy peers (29).

Parents of children with CF may perceive exercise more negatively than parents with healthy children (6). These concerns may be transmitted to the children who in turn begin to question their own ability to regularly participate in moderate-vigorous physical activities (60). It is possible that by improving their fitness levels, children with CF will be better able to cope physically and psychologically with performing vigorous activity, which in turn will maintain and improve their aerobic fitness levels.

A number of studies have examined the relation between indices of fitness and survival in CF patients. Nixon et al (61) found a significant relation between peak oxygen uptake (VO_2peak) and 8 year survival in children with CF. Patients with a VO_2peak ≥ 82% of predicted had an 83% chance of survival at 8 years. The 8 year survival rate for patients with a VO_2peak between 59-81% of predicted was 51%, and was only 28% in those with a VO_2peak ≤ 58% of predicted. CF patients with higher levels of aerobic fitness were 3 times more likely to survive than patients with lower aerobic fitness levels after adjusting for other risk factors including age, sex, body-mass index, FEV1 and end-tidal partial pressure of carbon dioxide at peak exercise.

Pianosi et al (66) measured pulmonary function and maximal oxygen uptake over a 5 year period in 28 CF patients between the age of 8 and 17 years. Initial VO_2peak was not predictive of 7-8 year survival. However, peak VO_2 fell by 2.1 $ml^{-1}kg^{-1}min^{-1}$ per year, and the rate of decline and final VO_2peak were significant predictors of 7-8 year survival. Patients with a peak VO_2 < 32.0 $ml^{-1}kg^{-1}min^{-1}$ had a dramatic increase in mortality compared to those with VO_2peak >45 $ml^{-1}kg^{-1}min^{-1}$. Arresting the decline in VO_2 could potentially increase the quality and length of life of CF patients. Klijn et al (39) found that changes in VO_2peak were related to changes in lung function and to a smaller extent changes in nutritional status in children with CF between the age of 4 and 19 years.

De Jong et al (13) examined the relation between pulmonary function, exercise capacity, dyspnoea and quality of life in 15 CF patients between the age of 16 and 40 years. Quality of life was assessed using the Sickness Impact Profile (SIP). There was a significant relation between overall SIP score and maximal exercise capacity. There was no relation between overall SIP score and FEV1 and FVC. In contrast, Orenstein et al (63) found that FEV1 was related to quality of life as measured by the Quality of Well-being scale.

Exercise Training

Aerobic training, anaerobic training, strength training or a combination may be beneficial to CF patients. Selvadurai et al (83) compared the effects of aerobic and resistance training on indices of health in children with CF. Activity levels, VO_2peak, and quality of life were higher following aerobic exercise training than resistance training. However, resistance training resulted in greater improvements in lung function, leg strength and weight gain than aerobic training. Orenstein et al (62) undertook a similar study in CF patients between the age of 8 and 18 years. A total of 67 patients were randomly assigned to a strength or aerobic training group. Body weight, strength and pulmonary function increased in both groups. The results indicate that strength and aerobic training are beneficial for health and well-being in CF populations.

Prescribing exercise to maintain and or improve indices of health and wellness in CF patients is a major challenge for both parents and allied health professionals. A recent study evaluated the effects of a 12 week medically supervised training programme on fitness and endurance performance in 12 sedentary CF patients (93). Subjects had a mean FEV1 of 71% and had no acute exacerbations in the previous 3 months. The training programme consisted of walking or running on a treadmill at 60% of maximum heart rate (HRmax) during the first 4 weeks. The training load was increased to 70% and 80% HRmax at week 4 and week 8 respectively. Exercise time, absolute and relative VO_2peak, and peak Ve increased in response to the training. There was no change in FEV1 or HRmax.

Although medically supervised exercise programmes allow for tailored and personalized training, they may not be attractive for large populations due to cost, time constraints and travel. There is emerging evidence that home-based physical activity programmes are a viable alternative to a medically supervised programme. Moorcroft et al (54) evaluated the effects of a one year unsupervised home based exercise programme in 48 adults with CF. Subjects were randomly assigned to a control group or an exercise group. The exercise group received personalised training aerobic training and resistance training programmes based on their exercise preferences. Subjects undertook three, 20 min sessions of upper body and 3 sessions of lower body exercises per week. They also kept a training diary and met with an exercise specialist every 4 weeks. A constant load submaximal leg ergometry and arm ergometry test was undertaken at baseline and at the end of the study. FEV1 declined by 67.0 ml and 174.0 ml in the exercise and control group respectively. The one year decline in FEV1 in the exercise group (1.5%) was lower than predicted (2-3 %). FVC improved by 46.0 ml in the exercise group and decreased by 167.0 ml in the control group. Blood lactate levels and heart rates were significantly lower in the exercise than the control group during the post training constant load submaximal leg ergometry test. There was no change in lactate concentrations and heart rates during the arm ergometer test.

Schneiderman-Walker et al (82) found a slower rate of decline in pulmonary function in 30 prepubescent and adolescent CF patients (7-19 yr) during a 3 year home-based aerobic exercise programme compared to a sedentary control group (35 CF patients). Although further training effects were not evident, the subjects in the exercise group reported positive attitudes and perceptual benefits from the training which in turn may improve motivation and adherence to exercise training. Improvements are only temporary and pulmonary function returns to pre-training levels within weeks of stopping the exercise programme (59).

Training programmes involving anaerobic activities provide variety and mimic many of the daily activities undertaken by children. Klijn et al (38) investigated the effects of a 12 week individually supervised anaerobic training programme in children (9-18 years) with CF. Patients were randomly assigned to a control (n=10) or training group (n=10). Training consisted of anaerobic activities lasting 20-30 sec. Each session was 30-45 min in duration, and was performed 2 d/wk. There were no within group or between group differences in body composition, pulmonary function, peripheral muscle force or habitual activity following the 12 week training programme. Mean power output, peak power output, maximal workload and VO_2peak increased at week 12 in the exercise group. The training group had a significantly higher score in the physical functioning domain of a quality of life questionnaire (HRQOL) than the control group. A large proportion (41%) of the change in the physical functioning was accounted for by the change in anaerobic peak power. At the end of the 12 week follow-up period, most of the measurements returned to pre-training levels except for absolute peak power output and the physical functioning domain of the quality of life questionnaire (38).

In summary, data from cross-sectional and longitudinal studies involving CF patients indicate that exercise training lowers mortality rates (65, 48) and improves pulmonary function, aerobic capacity, exercise tolerance, ventilatory muscle endurance, lung function, gaseous exchange (96), bone mass (44) and quality of life (52). Promoting physical activity should play an important role in medical management of CF.

Exercise Prescription

An exercise prescription can be defined as a specific guide provided to an individual in order to achieve his or her own measure of success or improvement. Exercise prescriptions can be general or specific, and can be applied to all of the health related components of fitness - cardiorespiratory endurance, skeletal muscle strength, skeletal muscle endurance, flexibility and body composition.

An exercise prescription should be physiologically effective and clinically safe (70). The exercise dose is usually characterized by the intensity, frequency, duration, progression and mode of exercise. The appropriate dose will depend on the desired outcome. For example, an improvement in resting blood pressure might be accomplished with an exercise intensity that is lower than that required to achieve an increase in VO_2peak. In addition, the frequency with which the exercise must be undertaken to have the desired effect varies with the intensity and duration of the session.

The intensity of exercise is usually based on physiological (heart rate, oxygen uptake, blood lactate), clinical (ECG abnormalities, angina pain and blood pressure abnormalities) and/or perceptual responses during a submaximal, or maximal exercise test. Physiologically based exercise prescriptions normally involve identifying an intensity range that requires a threshold level of oxygen consumption. Practical considerations make it difficult to measure VO_2 during exercise. However, the comparatively linear relation between HR and VO_2 for most dynamic exercises coupled with the fact that HR can be determined easily and accurately during exercise allows HR to be used for the purpose of exercise prescription. While the HR method of prescribing exercise is easy to calculate, it requires knowledge of age adjusted maximal HR, may be altered by emotional states and medication (16) and requires the acquisition of relatively expensive HR monitors that are often difficult to use and interpret, and may be uncomfortable to wear.

Rate of Perceived Exertion (RPE)

Exercise intensity can also be prescribed based on an individual's subjective impression of the overall effort, strain, discomfort or fatigue during the activity (68). The impression is referred to as a rating of perceived exertion (RPE). Perceptual responses interact and generally correlate with the physiological responses to exercise. RPE can be used to regulate exercise intensity for healthy children and adults across various exercise modes (18).

An individual's perception of physical exertion integrates sensory inputs between external stimuli arising from physical work and internal responses reflecting physiological functions and situational and dispositional factors (92). The physiological functions that mediate perceptual signals of exertion can be respiratory-metabolic, peripheral or non-specific. Sensations arising from the heart and lungs have been termed respiratory-metabolic or central signals of exertion, and are recorded as RPE-C (RPE-chest). RPE-C mediators include minute ventilation (Ve), respiratory rate (RR), oxygen consumption (VO_2), and carbon dioxide production (VCO_2) (47). Pulmonary ventilation and respiratory rate consistently account for a large proportion of the variation in rating of perceived exertion (RPE) during cycle ergometer exercise (46). Indeed, adjustments in respiratory rate during dynamic exercise appear to be one of the primary physiological mediators of respiratory metabolic signals of perceived exertion.

Peripheral or local signals refer to the contractile properties of the muscles (46) and include blood acidosis, blood glucose level, muscle blood flow and muscle fibre types. They are recorded as RPE-L (RPE-legs). The overall perceived exertion response (RPE-O) integrates the responses from the peripheral working muscles and joints, the central cardiovascular system, the respiratory system and the central nervous system (7).

Local factors are thought to be the most dominant influence during exercise involving small muscle groups. In contrast, central factors are more important in mediating perceptual ratings during exercise that involves the use of large muscle groups (94). At exercise intensities below 70% VO_2peak, the RPE for the active limbs (RPE-L) are higher than the RPE-C (47). This indicates that peripheral rather than local factors are the predominant sensory cues during exercise at low to moderate intensities. The local factors, which are reflected in RPE-C, do not appear to play a significant role until more vigorous exercise is performed (47). The RPE-O generally falls between the RPE-C and RPE-L. In individuals with pulmonary limitations such as chronic obstructive pulmonary disease or cystic fibrosis it is reasonable to expect that the RPE-C will be the dominant perceptual mediator.

A number of RPE scales have been developed to measure perceptual responses during exercise. The Borg 15-category scale is the most commonly used scale. It is comprised of numbers and verbal descriptors that represent the intensity of physical exertion (figure 1). The numerical scale values range from 6-20 and the associated verbal descriptors range from “fairly light” to “very, very hard”.

Rating of Perceived Exertion (RPE)	
6	No exertion at all
7	
	Extremely light
8	
9	Very light
10	
11	Light
12	
13	Somewhat hard
14	
15	Hard (heavy)
16	
17	Vey hard
18	
19	Extremely hard
20	Maximal exertion

Figure 1. Borg 15-Category Rating Scale of Perceived Exertion

Some children are unable to consistently assign words to numbers or phrases that represent exercise related feelings. Furthermore, many children are unable to understand or use the verbal cues that are not part of their current vocabulary. The OMNI scale was recently developed to overcome the methodological and semantic limitations that occur when children use adult scales such as the Borg 15-category scale to rate feelings of perceived exertion.

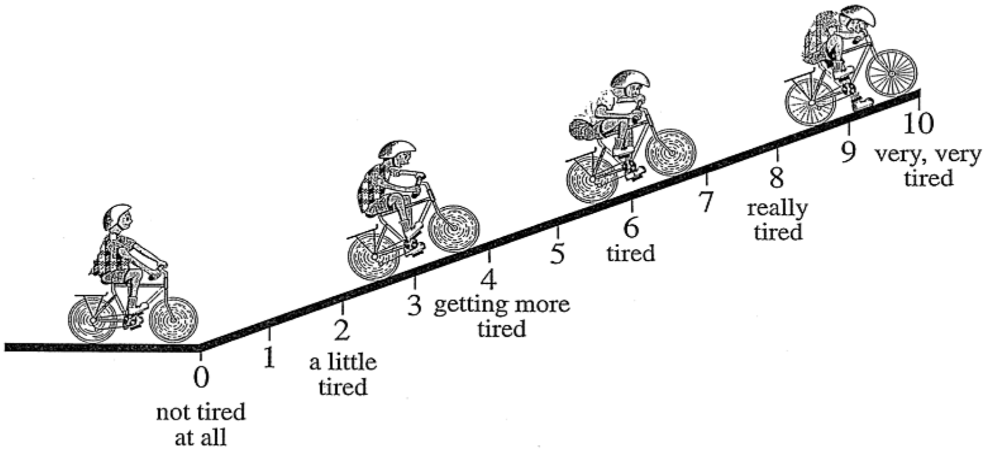

Figure 2. The OMNI Scale of Perceived Exertion: Child, Cycle Format

The OMNI Scale is comprised of verbal descriptors, pictures and a 0-10 point rating scale positioned along a visibly discernible hill in ascending order of perceived intensity (78). The scale (figure 2) was developed in four stages. Firstly, an artist drew 4 pictures illustrating a child experiencing various levels of exertion while cycling a bicycle up a hill. Children were then asked to verbally describe the intensity of physical effort portrayed in each picture. From the verbal descriptions provided by the children the word “tired” appeared frequently and was used as the base word to develop a scale of six verbal descriptors ranging from “not tired at all” to “very, very tired”. It is interesting to note that the words “hard” and “light” were not words frequently used by children to describe exertion even though these are the verbal descriptors found on the Borg-15 category scale.

RPE differentiated to the legs and chest (RPE-L and RPE-C) provide more precise information about anatomically regionalized mediators of effort perception. For example, RPE-L values are higher than RPE-C at the ventilatory threshold during cycling, and RPE-L has the largest mediating effect on overall RPE (RPE-O). The OMNI RPE value differentiated to the leg and chest (RPE-L and RPE-C) and undifferentiated or overall RPE (RPE-O) are distributed as positive linear function of VO_2 and heart rate among children of different race and gender (78). Robertson et al (76) confirmed that young boys and girls between the age of 8 - 12 years can discriminate between two intermittently presented target RPEs while self regulating cycle ergometer exercise (intensity discrimination). Children performed a graded maximal exercise test (estimation trial) and an exercise bout with a specified target RPE (production trial) on a cycle ergometer. RPE values were correlated with HR and VO_2. Heart rate and VO_2 values did not differ between estimation and production trials at the specified RPE value. The HR and VO_2 values were consistently higher when the children exercised at an RPE of 6 compared to an RPE of 2.

Perceptions of control during exercise are enhanced when exercise is self-selected (80) whereas exercise intensity prescription based on physiological parameters such as, VO_2 HR or lactate threshold may be perceived as highly controlling. In many instances, prescribed exercise may exceed an individual's preferred level of intensity and may establish negative attitudes toward physical activity contributing to low exercise adherence (42, 21). Longitudinal studies report that participants tend to deviate from prescribed levels of intensity in favour of their apparently preferred levels (81).

Allowing individuals to use effort perception to self select their preferred intensity may encourage the development of intrinsic motivation, a central element in promoting adherence to exercise, and increasing enjoyment and participation levels (97). Johnson et al (2006) found that 86% of women involved in aerobic exercise used effort perception exclusively to determine exercise intensity (34). This is not surprising considering that exertional feedback is often used unconsciously to regulate the pace of many daily activities. It also allows individuals adjust their work rate to account for improvements in fitness.

Providing exercise recommendations as opposed to exercise prescriptions helps move people away from the controlling culture that may establish negative attitudes to exercise (21). In a recent study, a group of overweight women were allowed to self-select their preferred exercise intensity on one occasion, and were prescribed a specific exercise intensity on another occasion. The exercise intensity did not differ more than 10%, but the level of enjoyment was significantly lower during the prescribed intensity exercise bout (21).

The majority of studies that have evaluated the use of effort perception to regulate exercise intensity have used walking. Spelman et al (87) covertly measured walking velocity during a typical walking session in 29 adult habitual walkers (22 women and 7 men) with a mean age of 35 yr. The subjects subsequently performed an 8-min level treadmill walk at a velocity determined in the typical walking session. The treadmill test was used to determine physiological, metabolic and perceptual responses at the self-selected pace. The mean walking speed was 1.78 $m \cdot s^{-1}$ and the effort was perceived to be “fairly light” (10.9) on the Borg 15-category scale. The average metabolic cost of walking at a self-regulated pace was 5.2 METS and corresponded to 52% VO_2max and 70% HRmax. Energy expenditure during a typical walking session was estimated to be approximately 257 kcal, or 3.8 $kcal \cdot kg^{-1}$. Using a similar design Murtagh et al (57) covertly observed the walking velocity in 11 women with a mean age of 40 years. The mean walking velocity corresponded to 59% VO_2max and 67% HRmax, and the mean RPE was 11.5 (fairly light). When asked to demonstrate their interpretation of "brisk" walking over the same course the velocity increased significantly to 1.79 $m \cdot s^{-1}$, which corresponded to 69% VO_2max, 79% HRmax and an RPE of 13.6. These findings indicate that when walking for exercise, adults self-select speeds and intensities that meet definitions of moderate intensity activity, and instructing them to walk briskly encourages vigorous exercise (57).

Porcari et al (69) evaluated the treadmill velocity among 17 men and 19 women (37.4 ± 5.1 years) during 40 min of treadmill walking at a self-selected intensity. The mean walking velocity was 1.7 ± 0.2 $m \cdot s^{-1}$ (3.9 mph). This corresponded to 44% VO_2max and 63% HRmax. The treadmill velocity was similar to that selected by the women in the Spelman et al study (1.78 mph). Lind et al. (2005) reported that %HRmax, %VO_2max, and RPE during the final 10 min of a 20 min walk at a preferred intensity in sedentary middle-aged women was 81%, 66% and 13.4 respectively. This intensity was accompanied by stable, positive ratings of affective valence (42).

Parfitt et al (65) compared the effects of 20 min of treadmill exercise at a prescribed intensity (65% VO_2max) and at a self-regulated intensity in 26 active college students (20.5 years) with an estimated VO_2max of 51.0 $ml \cdot kg^{-1} \cdot min^{-1}$. The mean self-regulated intensity was 71.0%VO_2max. Although the metabolic work rates differed between the prescribed and self-regulated exercise sessions, there was no difference in RPE, psychological affect or enjoyment between the two exercise sessions. In contrast, Glass et al (26) found that college students with an almost identical age and fitness profile to those studied by Parfitt et al, self-selected a treadmill walking intensity corresponding to 54% VO_2max.

Ekkekakis et al (20) and Lind et al (42) found that when individuals are allowed to self-regulate their exercise intensity, they will select an intensity that approximates the ventilatory breakpoint (Vpt). This is perhaps not surprising, considering that physical activity performed at or just below the Vpt will be perceived as enjoyable, and result in substantial health and fitness benefits. An intensity that significantly exceeds the Vpt precludes the maintenance of a physiological steady state, results in a systematic decrease in self-ratings of pleasure and is likely to be perceived as aversive (19). It is also possible that the negative affective response to a bout of exercise above the Vpt may, over time lead to aversion and avoidance of regular physical activity participation.

Lind et al (42) examined the intensity of physical activity that sedentary middle-aged women select during 20 min of treadmill exercise. They found that the %HRmax, %VO_2max, and RPE during a 20 min walk was similar to the intensity corresponding to the Vpt. The RPE values ranged from 12-14 on the 15-point Borg RPE Scale, which is similar to the RPE values associated with the Vpt reported in other studies (45, 87). The ratings of affective valence (decreases in pleasure or increases in displeasure) remained unchanged, and were within the positive (or nonnegative) range. The mean rating at each time point was not different from the affective valence associated with the Vpt.

Specific RPE ranges to target aerobic fitness improvements can be identified through response normalised modeling techniques. Response normalised modeling identifies a stable RPE or RPE range that corresponds to an exercise intensity that produces a specific physiological and/or psychological outcome and is also common to a specified and defined individual or group (77).

Situational factors that may influence the intensity of exertional perceptions include time, environmental setting and exercise mode. In contrast, dispositional factors are psychological traits that can predictably shape a person's response over a wide range of settings (92). An individual's exercise intensity depends in part on his or her affective response (pleasure or displeasure) resulting from sensory experiences from various parts of the body. Affect refers to a short duration, high intensity emotion or feeling. When given a choice, individuals will generally adjust their effort intensity during exercise to maximize affect. The short term rewards of positive affective responses that can be derived from a positive exercise experience might assist people to stay motivated to sustain regular physical activity and avoid drop out.

Affect is also thought to be one of the psychological factors contributing to RPE. Hardy and Rajeski (30) found a moderate relation between RPE and affect score at various exercise intensities, with stronger correlations at higher and lower intensities than at moderate intensity exercise. Cabanac (8) examined the effect of allowing subjects to self-regulate speed or grade during treadmill exercise. Healthy young adult men were given the task to climb a 300 m elevation on a treadmill, at 5 X 5 combinations of speed and grade. If the grade was imposed and speed self adjusted, or speed imposed and grade self adjusted, the men spontaneously climbed the 300 m in a constant time due to reciprocal adjustments in speed and grade that resulted in the maintenance of an approximately constant power. The individual choices could be predicted from the sum of ratings for displeasure in the chest and lower limbs. The authors speculated that the affect resulting from sensory inputs from the chest and limb muscles were responsible for optimizing muscular work.

It is likely that exercise can be sustained as an enjoyable experience if individuals can select one or more activities that are perceptually pleasing and maintains affective valence. A prescription that exceeds an individual's preferred level of intensity may lead to a reduction in affect during exercise. Sheppard et al (84) tracked the temporal dynamics of affective responses during and after exercise at different intensities in young adolescent boys and girls with a mean age of 13.3 years. Participants cycled for 15 min at a low intensity (80% of ventilatory threshold power output), high intensity (130% of ventilatory threshold power output) and a self-regulated intensity. The mean workrate was 121 ± 21 W, 75 ± 15 W, and 83 ± 22 W, for the high intensity, low intensity and self-regulated condition respectively. Affective valence assessed using the Feeling Scale was positive during exercise in the low intensity and self-regulated condition and declined in the high intensity condition. However, the divergent trends between intensity conditions were diminished post exercise due to a rebound in response in the high-intensity condition. Solomon (86) proposed the opponent-process theory to explain this rebound effect. The theory implies that if the exercise stimulus is perceived as aversive, then the affective state in the post exercise condition would be relatively positive. Conversely, the affective state during recovery could be relatively negative if the work stimulus was perceived as enjoyable and the cessation of exercise was seen as a "letdown".

CHAPTER III

METHODOLOGY

Subjects

Twelve children with CF volunteered to participate in the study. Characteristics of the subjects are presented in table 1. CF was diagnosed on the basis of a positive sweat test or pulmonary function assessment. The subjects were familiar with the pulmonary function and exercise testing procedures, and demonstrated cognitive ability to read each verbal descriptor on the Children's OMNI scale. The study was approved by the Research Ethics Committee at Dublin City University, and the Scientific and Ethics Committee at Temple Street Children's University Hospital.

Experimental Design

Subjects made 3 separate visits to the Human Performance Laboratory (HPL) in the School of Health and Human Performance at Dublin City University. Each visit was approximately 1-2 h, and was separated by a minimum of 72 h. Subjects were requested to fast for 4 h and refrain from strenuous physical activity for 24 h prior to each visit. The first visit was used to obtain assent/consent, determine VO_2peak, and anchor the Children's OMNI scale. The second visit was used to familiarize children with the use of the Children's OMNI scale. During the final visit subjects exercised for 20 min on a cycle ergometer. The exercise intensity was self-regulated.

Screening

Participants provided written assent and their parents/guardians consented to their participation, after being fully informed of the nature of the research. Subjects subsequently underwent a brief physical examination, had their height, weight and body composition measured, performed a spirometry test to assess pulmonary function and a maximal exercise test to measure aerobic capacity.

The maximal exercise test was performed on an electronically braked cycle ergometer (Ergoline, COSMED, Rome, Italy) using a ramp protocol. Following a 2 min warm-up at 5-10 W, the resistance was increased by 5-10 W every min until the subject reached volitional fatigue. Subjects were verbally encouraged to give their best effort. Breath by breath respiratory, metabolic and gas exchange variables were continuously measured and heart rate (HR) was recorded every min.

The low and high perceptual anchors for the OMNI Scale were established using a visual interface cognitive procedure in conjunction with the maximal exercise test. This procedure required the subject to cognitively establish a perceived intensity of exertion that was consonant with that depicted visually by the cyclist at the bottom (i.e., low anchor, rating 0, at the beginning of exercise) and top (i.e., high anchor, rating 10, at the end of the maximal exercise test) of the hill as presented in the OMNI Scale illustrations.

Familiarization Trial

Subjects performed three, 6-min bouts of sub maximal exercise at 40%, 60% and 80% VO_2peak. The workloads were presented in a randomized order and recovery between bouts was sufficient to allow the HR return to resting values. Respiratory metabolic responses were continuously measured, and HR and RPE were recorded every min and every second minute respectively, during each test.

Experimental Trial

During the final visit, subjects exercised for 20 min at their preferred intensity on an electronically braked cycle ergometer. They were given the opportunity to change the resistance every 5 min. This was achieved using hand gestures. The subjects indicated to the tester whether they wanted the intensity to be increased, decreased or kept constant. Respiratory metabolic and gas exchange variables were measured continuously using open circuit spirometry (SenorMedics, Yorba Linda, CA). Heart rate and RPE were recorded every min and every second min respectively during the test.

Laboratory Procedures

Anthropometrics

Body weight (kg) and height (cm) were determined without shoes using a Seca scale and attached stadiometer (Seca 220, Telescopic Measuring Rod, Vogel and Halke, Hamburg, Germany)

Heart Rate

Heart rate was continuously measured using a 4 lead ECG system (Sensormedics, Yorba Linda, CA). Four electrodes were placed on the chest prior to each test to facilitate continuous ECG monitoring.

Rating of Perceived Exertion

RPE was measured using the Children's OMNI scale (78). An undifferentiated rating was estimated for the overall body (RPE-O), and differentiated rating was estimated for peripheral perceptions of exertion in the legs (RPE-L), and respiratory-metabolic perceptions in the chest (RPE-C). A definition of perceived exertion appropriate for children and an explanation of the OMNI scale and instructions on how to use the scale to rate perceived exertion were read to the subjects prior to commencing the tests (appendix II).

Pulmonary Function Assessment

Forced expiratory volume in one second (FEV_1), forced vital capacity (FVC), total lung capacity (TLC) and maximal breathing capacity at rest (MVV) were determined using a spirometry system (Sensormedics Vmax 229, Loma Linda, CA, USA). A mass flow sensor coupled to a disposable mouthpiece was used to collect measurements of ventilation. The mass flow sensor was calibrated prior to each test using a 3 L volume syringe (Sensormedics, Loma Linda, CA, USA).

Cardiorespiratory and Metabolic Measurements

Expired gases and ventilatory volume were determined using a Sensormedics Vmax 229 metabolic system. The gas analysers were calibrated prior to testing with standard gases of known concentration. A mass flow sensor (Sensormedics, Loma Linda, CA, USA) was used to collect breath-by-breath measurements of ventilation. The mass flow sensor was calibrated prior to each test using a 3 L volume syringe (Sensormedics, Loma Linda, CA, USA).

The mass flow sensor is a low resistance tube with a tapered internal diameter extending from both ends of a laminar flow throat. A cold and hot stainless steel wire electrically heated to -180^{o} and -240^{o} respectively, are centered in the flow stream. These wires are elements in a servo-controller bridge circuit that maintains the resistance ratio of the two wires at a constant value. If only the temperature of the inspired gases change, then both wires lose heat at the same rate and no current change is required to keep the bridge balanced. As air flows across the wires, the hot wire loses heat more rapidly than the cold wire and the current must be added to keep the bridges balanced at a 3:4 ratio. The amount of current required is proportional to the mass flow of the gas. This method ensures that the sensor measures only the heat loss from the molecular convection of the moving gas stream, and not the artifact due to cooling of the gas as it passes through a breathing assembly.

The mass flow meter responds to instantaneous flow rates between 0-16 $L\cdot sec^{-1}$ and integrated flow between 0-350 $L\cdot min^{-1}$ with flow resistance <1.5 $cmH_2O\cdot litre^{-1}sec^{-1}$. The mass flow sensor was outputted to the analyser module of the Vmax 229 and was sampled at a rate of 125 Hz.

Volume Calibration

A 3 L volume syringe (Sensormedics, Loma Linda, CA, USA) was used to calibrate the mass flow sensor. The syringe was connected to the mass flow sensor, and stroked four times in order to measure inspired and expired volumes. The volumes were calculated by expressing 3 L as a fraction of each measured inspired and expired volume achieved during calibration. An average correction factor was then calculated for inspired and expired volumes, and used to fine-tune the volume measurement. A verification procedure was performed by stroking the 3 L volume syringe 4 times. Inspired and expired volumes were measured using the newly calculated correction factors. In order to pass the calibration procedure one of the four strokes had to have an average flow rate <0.5 $L \cdot sec^{-1}$, and at least one of the four strokes had to have an average flow > 3.0 $L \cdot sec^{-1}$.

Gas Analysers - Calibration

The Vmax 229 utilizes a rapid response non-dispersive infrared measurement technique. An O_2 and CO_2 analyser is integrated within the Vmax 229. A small sample of inspired air is drawn through a sample cell, and exposed to an infrared light through an optical that is passed through a band-pass filter and the sample cell. An infrared detector responds to the amount of infrared light that passes through the sample cell. Carbon dioxide absorbs infrared light over wavelengths ranging from 0.7 μm to 15 μm. The amount of light passing through the sample cell varies according to the concentration of CO_2 in the sample cell. Based on measured levels of infrared light intensity, the analyser computes the PCO_2 in the gas sample. The CO_2 analyser is linearly scaled across the 0-100% range with a resolution of 0.01 %CO_2, and a response time of <130ms (10-90%) at 500 $ml \cdot min^{-1}$ flow.

The O_2 analyser is based on the high paramagnetic susceptibility of O_2. A diamagnetic glass dumbbell suspended in a magnetic field rotates in proportion to the PO_2. The analyser is linearly scaled across the 0-100% range with a resolution of 0.01 %O_2 and a response time of <130 $m \cdot sec^{-1}$ (10-90%) at 500 $ml \cdot min^{-1}$ flow.

The gas analysers were calibrated using gases of known concentration (BOC gases, Dublin, Ireland). The gas composition in tank 1 was 26.00 ± 0.02 %O_2 and the balance N_2. The gas composition in tank 2 was 4.00 ± 0.02 %CO_2, 16.00 ± 0.02 %O_2 and the balance N_2. A small bore drying tube connected to the CO_2 and O_2 analysers samples the calibration gases and inhaled/exhaled air. The absorption and evaporative properties of the drying tube ensured that the relative humidity in the inhaled/exhaled gas and in the calibration gas was equilibrated to ambient conditions before being samples by the analysers.. Samples of both calibration gas and gas inhaled by subjects were taken at a rate of 125 Hz. The response time of the CO_2 analyser was synchronised with that of the O_2 analyser.

Lactate and Ventilatory Threshold Determination

The ventilatory breakpoint (Vpt) was determined using the V-slope method (5). This procedure involves plotting CO_2 against O_2, and a line parallel to the line of identity is plotted through the lower data points. This procedure identifies the point during the exercise test where CO_2 begins to rise disproportionately faster than O_2, due to the bicarbonate buffering of the lactic acidosis.

Statistical Analysis

Descriptive statistics for perceptual and physiological variables were calculated as mean ± SD. A one-way repeated measures ANOVA was used to compare perceptual and physiological variables during the 20 min self regulated cycle ergometer exercise. Significant main effects were probed using the Tukey post-hoc analysis. A paired sample t-test was used to compare the mean physiological responses at Vpt and during the self-regulated cycle ergometer exercise. SPSS for Windows statistical software was used to perform the statistical analysis. Statistical significance was accepted at the $P < 0.05$ level of confidence

CHAPTER IV

RESULTS

Subject characteristics are presented in Table 1, and the physiological responses at peak exercise are listed in table 2. Physiological and perceptual responses for each 5 min interval during the 20 min self-regulated cycling exercise are presented in table 3. Undifferentiated RPE (RPE-O) and differentiated RPE for the chest (RPC-C) and legs (RPE-L) were lower at 5 min compared to 10 min, 15 min and 20 min. Both absolute and relative VO_2, heart rate and work-rate remained stable throughout the 20 min exercise bout.

Perceptual and physiological responses did not change during the final 15 min of exercise. The average value during this time period was calculated and used to determine the responses during self-regulated exercise. The mean work-rate was 24.7 W, and the RPE-O, RPE-C and RPE -L were 5.8 ± 2.6, 4.5 ± 2.6 and 5.7 ± 2.4 respectively. Exercise intensity, expressed as $\%VO_2$peak and % heart rate max (%HRM) was 53.2 ± 10.9 and 73.1 ± 8.5 respectively. The mean MET value and caloric expenditure (Kcal/min) were 6.4 and 4.7 respectively

The $\%VO_2$peak and %HR peak at the Vpt was 63% and 82% respectively. The mean VO_2 during self-regulated exercise was lower ($p<0.001$) than the mean VO_2@Vpt (Figure 3). The mean HR during self-regulated exercise was also lower ($p<0.001$) than the mean HR@Vpt (Figure 4). There was a linear relation between VO_2peak and pulmonary function (FEV1, $r=0.70$ $p<0.01$; FVC $r=0.90$, $p<0.001$, MVV $r=0.70$ $p<0.01$) in children with CF.

Table 1: Subject characteristics

Subject	Age (yr)	Height (cm)	Weight (kg)	BMI ($kg \cdot m^{-2}$)	SBP (mmHg)	DBP (mmHg)	%FEV1 (predicted)	%FVC (predicted)	FEF (25-75%)
1	13	161.0	49.5	19.1	126	67	72	87	27
2	10	139.0	41.8	21.6			127	127	79
3	10	151.0	34.7	15.0	104	80	64	67	34
4	10	139.0	30.5	15.8	117	60	99	104	42
5	12	143.0	36.3	17.8	132	82	95	109	34
6	15	145.5	44.4	21.0	118	74	52	92	11
7	9	127.0	26.1	16.2	100	53	74	96	20
8	11	145.5	44.2	20.9	117	72	70	76	37
9	13	145.4	36.9	17.5	101	64	63	75	19
10	9	128.0	23.3	14.2	106	80	106	98	55
11	10	134.5	27.7	15.3	112	84	65	82	15
12	8	129.0	24.4	14.7	110	73	99	99	41
Mean ± SD	10.8 ± 2.0	140.7 ± 10.1	35.0 ± 8.7	17.4 ± 2.6	113.0 ± 10.2	71.7 ± 9.8	82.2 ± 22.2	92.7 ± 16.6	34.5 ± 18.9
Range	8-15	127-161	23.3-49.5	14.2-21.6	100-132	53-84	52-127	67-127	11-79

SBP, systolic blood pressure, DBP, diastolic blood pressure, BMI, Body mass index

Table 2: Physiological responses at peak exercise

Subject	VO_2 ($ml^{-1} \cdot kg^{-1} \cdot min^{-1}$)	VO_2 ($L \cdot min^{-1}$)	Heart rate ($b \cdot min^{-1}$)	Work rate (Watts)	Ventilation ($L \cdot min^{-1}$)
1	41.03	2.03	187	115	58.83
2	33.73	1.41	181	80	37.50
3	42.50	1.47	175	90	39.33
4	56.17	1.71	181	90	38.10
5	43.03	1.56	177	85	43.17
6	25.95	1.15	174	55	33.85
7	39.30	1.02	180	60	30.20
8	27.23	1.20	156	85	23.80
9	37.47	1.38	179	85	37.33
10	47.35	1.10	191	60	28.95
11	32.23	0.89	177	55	28.87
12	44.53	1.08	165	55	26.87
Mean ± SD	39.21 ± 8.59	1.34 ± 0.33	176.92 ± 9.24	76.25 ± 19.08	35.57 ± 9.3
Range	25.95-56.17	0.9-2.0	156-191	55-115	23.80-58.8

Table 3: Perceptual and physiological responses during self-regulated cycling exercise

	Time (min)			
Variable	**0-5**	**5-10**	**10-15**	**15-20**
RPE-O	2.67 ± 2.23†	5.00 ± 3.31	5.38 ± 3.06	5.68 ± 3.15
RPE-C	1.75 ± 1.83†	3.46 ± 2.78	4.36 ± 2.99	4.60 ± 3.19
RPE-L	2.38 ± 1.90†	4.43 ± 3.01	5.64 ± 2.99	5.64 ± 3.14
VO_2 ($l \cdot min^{-1}$)	0.68 ± 0.22	0.74 ± 0.24	0.68 ± 0.17	0.73 ± 0.23
VO_2 ($ml^{-1} \cdot kg^{-1} \cdot min^{-1}$)	20.55 ± 7.52	25.81 ± 16.03	19.99 ± 4.68	20.96 ± 4.85
$\%VO_2$	51.65 ± 14.93	56.99 ± 15.84	51.57 ± 10.25	54.08 ± 10.44
Heart rate (bpm)	124.78 ± 14.60	131.05 ± 17.15	126.65 ± 13.37	129.20 ± 11.84
% Heart rate	70.77 ± 9.31	74.30 ± 10.44	71.86 ± 9.20	73.17 ± 7.19
Workrate (watts)	25.00 ± 15.95	27.92 ± 18.02	20.42 ± 11.37	25.83 ± 16.90
% Workrate	33.11 ± 19.16	37.07 ± 21.35	25.79 ± 10.85	31.66 ± 16.02

†P<0.001 vs 5- 10, min 10-15 min 15-20 min†

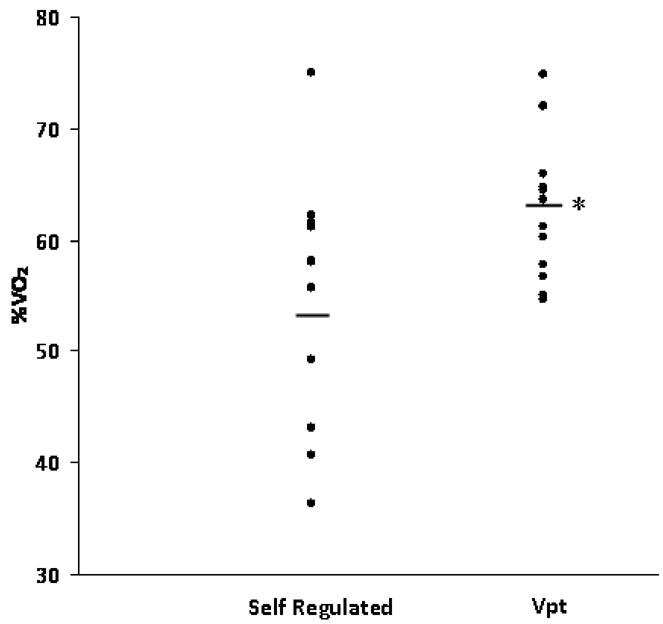

Figure 3: Individual and mean %VO_2peak values during self-regulated exercise and at the Vpt. * p<0.001 vs. self-regulated

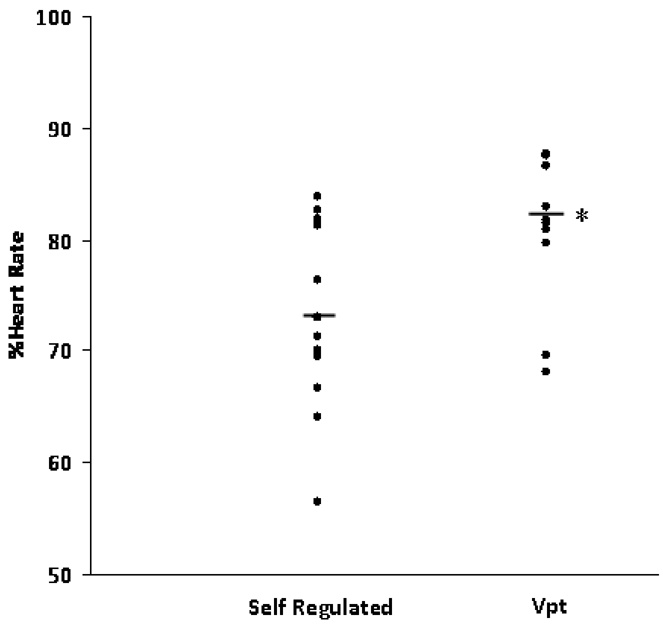

Figure 4: Individual and mean %HRpeak values during self-regulated exercise and at the Vpt. * p<0.001 vs self regulated

Chapter V

DISCUSSION

The purpose of this study was to assess the physiological and perceptual responses during 20 min of self-regulated exercise in children with CF. The OMNI scale of perceived exertion was used to assess ratings of perceived exertion (RPE). The OMNI scale is a pictorial scale that was developed to overcome the methodological and semantic limitations that occur when children use RPE scales designed for adult use.

Physiological Characteristics

Height, weight and BMI were on average, similar to values reported for healthy children of the same age (100). However, when each of the participants were compared to age specific norms, 5 children were classified below the 25th percentile for height, 6 were between the 25th-75th percentile and 1 child was above the 75th percentile. There were similar findings for both weight and BMI scores; 4 children below the 25th percentile, 6 between the 25th-75th percentile and 2 above the 75th percentile.

Respiratory Measurements

Forced expiratory volume in one second (FEV1) measures the volume of exhaled air during the first second of expiration. A comparison of actual FEV1 values with normal predicted values is commonly used to establish the severity of lung disease. Average FEV1 values for the children with CF were 15.2% and 16.5% below normative values for healthy boys and girls respectively (regression equations of Rosenthal, 1993, cited in (96) Wagner 1996). Values ranged from 27% above to 48% below normative values. A total of 4 boys and

4 girls had an FEV1 value below the predicted value for their age and height. Only 2 of the subjects had FEV1 values higher than normative values.

Forced vital capacity (FVC) is the maximal volume of air that can be forcibly expired following maximal inspiration, and is a measure of large airway patency. The maximal mid-expiratory flow rate (MMEFR) is the average forced expiratory flow between 25% and 75% (FEF 25-75%) of FVC manoeuvre. It is regarded as a more sensitive measure of small airway narrowing than FEV1. The average FEF 25-75% values for the children with CF were 40% and 36% lower than the norm values for healthy age and height matched boys and girls respectively. Although average FEF 25-75% values indicate mild small airway obstruction (50-60% of predicted), 3 girls and 1 boy had moderate obstruction (35-49% of predicted), and 2 girls and 2 boys had normal values.

The progressive loss of pulmonary function in CF patients is normally associated with a decrease in exercise tolerance which is manifested in a low VO_2peak. Similar to other studies (59), we found a significant positive linear relation between VO_2peak and FEV1 (r^2=0.78) and between VO_2peak and FVC (r^2=0.84) in children with CF.

Aerobic Capacity

VO_2peak values expressed in absolute terms and relative to body mass, were lower in the CF children than values reported for healthy children of a similar age (3, 4, 24). Mahon et al (1998) examined the physiological and perceptual responses in healthy 10 yr old boys during exercise on a cycle ergometer. The absolute and relative VO_2peak values were approximately 19% and 21% higher respectively, than the children in the present study (47).

A lower than normal reference value for fitness in children with CF (44) is a cause for concern considering that aerobic fitness (61) and the rate of decline in VO_2peak (66) are important predictors of survival in CF patients.

We did not measure levels or patterns of physical activity. Studies have shown comparable levels of weekday physical activity between healthy children, and children and adolescents with CF (60, 44). Nixon et al (60) used the Kriska Modifiable Activity Questionnaire (MAQ) to measure physical activity in 30 healthy subjects and 30 patients with CF between the ages of 7-17 years. Although the patients with CF were as active as their healthy peers they engaged in less vigorous physical activities. The reduced time spent in vigorous physical activities could not be explained by the extent of lung disease, aerobic fitness or nutritional status for the total group of 30 CF patients. However, when the 10 children with the lowest lung function were examined separately, vigorous and MET-hours of activity were significantly related to pulmonary function and aerobic fitness. It is worth noting that the majority of the children in the present study were encouraged to participate in physical activity and were involved in a minimum of one after school sports activity.

Self-Regulated Exercise Intensity

An exercise prescription is characterised by the intensity, frequency, duration, progression and mode of exercise. Exercise intensity is a crucial component of an exercise prescription and has a major influence on the extent to which exercise participation can lead to health and fitness benefits. Depending on the desired outcome, the intensity of exercise is based on the relative force of muscle action and/or the relative stress placed on the metabolic and cardiorespiratory systems. Exercise prescriptions designed to maintain or improve cardiorespiratory fitness are commonly based on one or more physiological measurement such as heart rate, VO_2 and/or blood lactate levels. An exercise prescription based on one or more physiological measurements may exceed an individuals preferred level of intensity, and may establish a negative attitude toward physical activity.

In a recent study, Johnson et al (34) interviewed a group of women involved in regular aerobic exercise. The women exercised for approximately 45 min per session on 4 - 5 days per week. A large proportion (84%) of the women used effort perception exclusively to determine exercise intensity. In another study (20), a group of overweight women were allowed to self-regulate their exercise intensity on one occasion, and were prescribed a specific exercise intensity, on another occasion. The exercise intensity did not differ more than 10% between the exercise bouts, but the level of enjoyment was significantly higher during the self-regulated exercise bout than during the prescribed exercise bout. This may be due in part to the fact that perceptions of control are enhanced when exercise is self-regulated (80).

When children with CF were allowed to self-regulate exercise intensity during 20 min of cycle ergometry, they selected a mean intensity corresponding to 54% VO_2peak and 73% HRmax. These values are within the moderate to vigorous (40-85% VO_2 peak) exercise intensity range advocated for health and fitness benefits (1). Although the majority of children selected a workrate between 50-60% VO_2peak, there was a large intra individual variation (Figure 5). One subject selected an intensity >70% VO_2peak while three others selected an intensity ≤45% VO_2peak. The average metabolic cost of cycling at a self-regulated pace was 6.4 METS and corresponded to a caloric expenditure of 4.7 $kcal \cdot min^{-1}$.

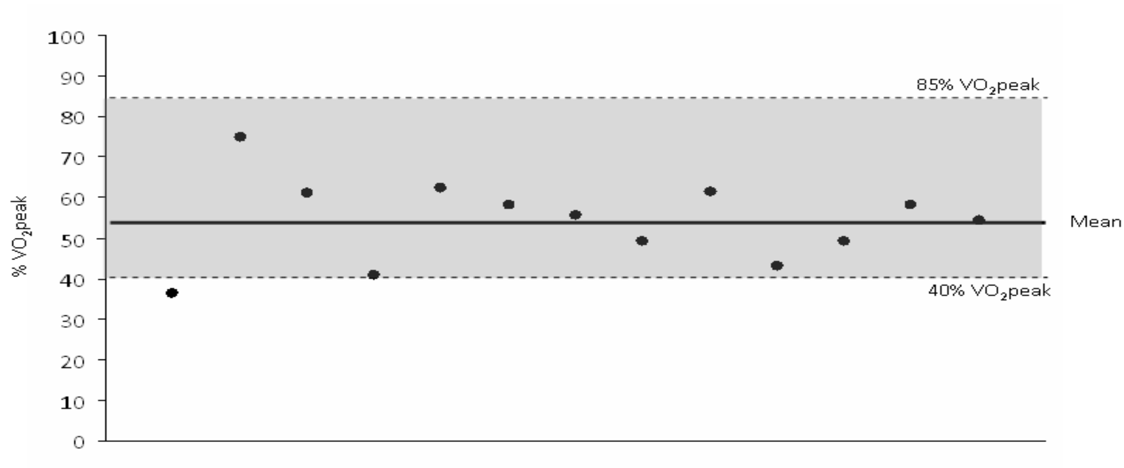

Figure 5: Mean %VO_2peak for each participant during the final 15 min of self-regulated exercise. Shaded area indicates the moderate to vigorous exercise intensity range advocated for health and fitness benefit.

To our knowledge no previous studies have examined the physiological and perceptual responses during self-regulated cycling in healthy children or children with CF. Dishman et al (1994) compared RPE, state anxiety, %VO_2peak, %Vpt, and blood lactate concentration in 11 high-active and 12 low-active men (23 ± 3 yr) at self-regulated power outputs during 20 min of cycling. Although the high-active group selected higher power outputs than the low-active group, both groups cycled at an intensity that equated to 62% VO_2peak (17).

The majority of studies that have examined the physiological and perceptual responses during self-regulated exercise in healthy adults have involved walking. Spelman et al (87) measured the walking velocity during a typical outdoor walking session in 29 healthy, adult habitual walkers. The subjects self-regulated their intensity, and subsequently performed an 8-min level treadmill walk at a velocity determined in the typical walking session. The mean selected intensity during a typical walking session (52% VO_2max and 70% HRmax) was almost identical to the values for children with CF in the present study. Others have reported similar metabolic and cardiovascular responses in middle aged women (42), active college students (26), overweight women (67), low-active and high-active men (16) and recreational walkers (57) when allowed to self regulate their walking intensity.

Rate of Perceived Exertion

The mean RPE-O during self-regulated exercise was 5.8, which corresponds to the verbal descriptor "tired". Differentiated and undifferentiated RPE values increased during the first 5 min of exercise and then remained constant. In contrast, heart rate and VO_2 values were similar throughout the 20 min of exercise. It is possible that central and peripheral signals mediating perceived exertion need time to adjust during steady state exercise. Lind et al (2005) found that speed, lactate, VO_2 and HR values did not reach steady state for 5-10 min during 20 min of self-regulated treadmill exercise in sedentary middle aged women (42).

Differentiated RPE's arising from the legs and chest reflect peripheral signals and respiratory metabolic signals respectively (7). In the present study the children with CF reported higher values for RPE-L than RPE-C during self-regulated exercise. The fact that signals of exertion arising from the legs appears to dominate the RPE response was surprising considering that CF patients normally compensate for the increased dead space associated with pulmonary function deterioration by utilizing a higher minute ventilation for a given VO_2 (10). The higher ventilation may be associated with a greater sense of breathlessness (37), and a higher than normal RPE-C value would consequently be expected.

The higher differentiated RPE arising from the legs than the chest may be due in part to disease severity and mode of exercise. Since none of the subjects has severe airway obstruction, it is unlikely that minute ventilation would have been altered to any great extent during exercise. Cycling is not a common activity among children in Ireland. Only 1% and 7% of 15-17 year old Irish girls and boys, respectively, cycle to post primary school (58). The proportion of fifth and sixth class primary school children who cycle to school is 1% (99). It is possible therefore that peripheral physiological mediators arising from untrained muscles in the legs increase the perception of effort.

Children are able to use the words and pictures of the OMNI scale to translate into numbers their perceptions of physical exertion (72, 73, 78, 74, 75, 76, 79, 89, 95, 28), and subsequently use these scales to self-regulate exercise intensity (76). Using an estimation production paradigm Robertson et al (2002) reported that 8-12 year old children were able to reproduce specific VO_2 values using the OMNI RPE scale as a guide. The children were able to reproduce RPE values during cycling exercise indicating that self-regulation of exercise using the OMNI scale as a guide is possible among a group of healthy male and female children (76). However, among our CF cohort, we found a wide variability in the perceptual response range during self-regulated exercise (Figures 6-8). This cannot be explained by disease severity as all but one subject had mild CF.

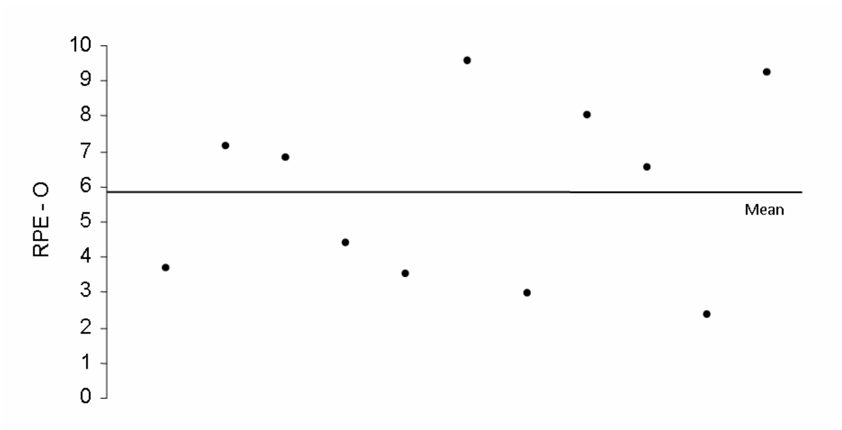

Figure 6: Mean individual RPE-O values during self-regulated exercise.

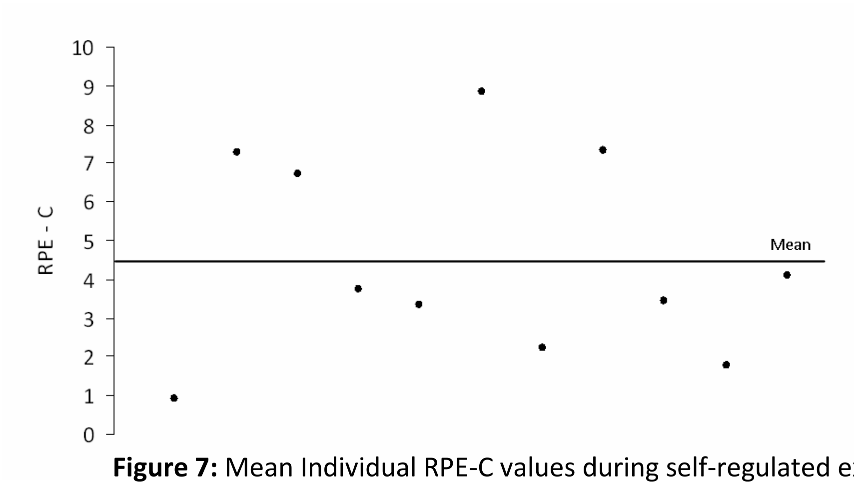

Figure 7: Mean Individual RPE-C values during self-regulated exercise.

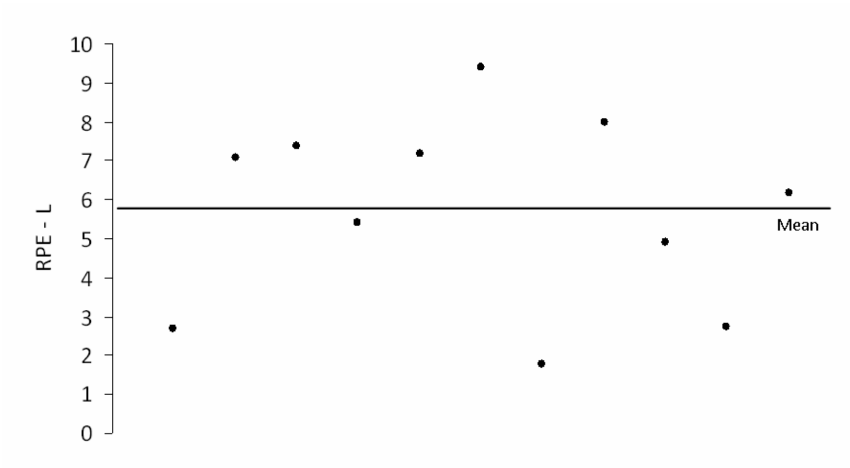

Figure 8: Mean individual RPE-L values during self-regulated exercise.

Ventilatory Breakpoint

There is a general consensus that the Vpt indicates the point of transition from aerobic to anaerobic metabolism during exercise (20). The %HRmax, $\%VO_2max$ and RPE response range that span the Vpt appear to be independent of gender (15), fitness (33) and exercise mode (32). Although some studies have reported increases in ventilation above Vpt during exercise to be independent of increases in blood lactate concentration, the majority of investigators have shown that changes in ventilation are accompanied by changes in blood lactate response to exercise (68). Exercise undertaken above the Vpt may be uncomfortable and difficult to maintain due to an accumulation of lactic acid. When allowed to self-regulate exercise intensity healthy adults select an intensity that approximates Vpt or LT (19).

The %VO_2@Vpt was 63%, and is similar to that reported previously by Mahon et al (1998) for healthy 10 yr old boys (67.2 ± 3.5%)(44) and healthy 11 yr old boys and girls (65%) (47). Anderson and Mahon (2) found a similar value (68%) in healthy 11 yr old boys. Although not reported, the workrate corresponding to the Vpt is probably higher in healthy children due to the fact that they generally have a higher VO_2peak than children with CF.

We hypothesized that when allowed to self-regulate exercise intensity during cycle ergometry, the majority of children with CF would exercise within ±10% VO_2@Vpt. The mean exercise intensity expressed as a %VO_2peak (53%) was 10% below the mean %VO_2@Vpt (63%). However, when analysed by individual subject, the majority (n=8) selected an exercise intensity ±20% VO_2@Vpt, and 4 subjects selected an intensity that was 31 to 38% below the VO_2@Vpt (Figure 9).

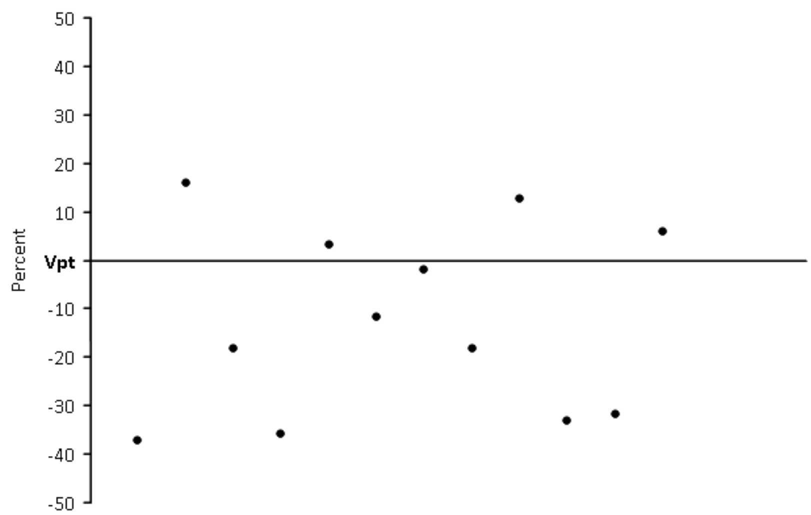

Figure 9: Percent of Vpt for each subject during self-regulated exercise

The RPE-O corresponding to the Vpt was not determined in the present study. However, the mean RPE-O of 5.8 was almost identical to the reported RPE-O value (6.0) at the Vpt in children with average and above average aerobic fitness levels (77). Although there were large intra-individual variations in the exercise intensity selected, the present findings suggest, that when allowed to self-regulate their exercise intensity children with CF will exercise at an intensity ≤ $\%VO_2@Vpt$ and at RPE 6 (tired) on the OMNI scale.

Limitations of the study

The majority of subjects had mild CF making it difficult to apply the results of the study to children with more severe CF. Disease severity increases with age so it is expected that the children will develop moderate or severe CF later in life. Due to the low severity of the disease in the study cohort, pulmonary limitations to exercise were not as evident as seen by the fact that the RPE-L values were higher than the RPE-C values in the self-regulated exercise sessions. Cycling is not a popular exercise modality among Irish children. Treadmill walking and/or running may have been a more appropriate exercise modality for investigating physiological and perceptual responses. Finally, the continuous nature of the exercise may not accurately reflect the intermittent nature of children's play.

Conclusion

Aerobic fitness and the rate of decline in VO_2peak are important predictors of survival in CF patients (61, 66). The mean VO_2peak for children in the present study was approximately 20% below values reported for healthy children. Children with CF were able to self-regulate their exercise intensity as evident by a stable heart rate, VO_2 and RPE.

Although the mean %VO_2peak was within the moderate to vigorous intensity range recommended for health and fitness benefits, there were large intra-individual variations in RPE, %VO_2peak, %HRpeak and %Vpt. Exercise prescription for children with CF should be individually tailored, and should take account of RPE in addition to conventional physiological indices of relative metabolic intensity.

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Appendix I

Table 4: Individual RPE-O values during self-regulated exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1				
2	3	5	2	4
3	2	4	8	10
4	2	5	7	9
5	2	3	5	6
6	2	3	4	4
7	5	9	10	10
8	1	2	5	3
9	6	10	8	6
10	3	8	6	6
11	1	2	3	3
12	7	9	10	9

Table 5: Individual RPE-L values during self regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1				
2	2	3	2	3
3	2	4	8	10
4	3	6	8	9
5	2	4	6	7
6	3	6	8	8
7	5	9	10	10
8	1	0	4	1
9	7	10	8	6
10	2	5	5	4
11	1	2	3	4
12	3	5	8	6

Table 6: Individual RPE-C values during self regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1				
2	0	1	0	2
3	2	5	8	10
4	3	5	7	8
5	1	2	4	6
6	1	3	3	4
7	6	8	9	10
8	0	1	4	2
9	5	9	8	5
10	1	3	4	3
11	1	1	2	2
12	2	4	4	4

Table 7: Individual VO_2 ($L{\cdot}min^{-1}$) values during self regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1	0.502	0.476	0.717	1.020
2	0.845	1.203	0.854	1.112
3	0.915	0.946	0.838	0.920
4	0.674	0.744	0.598	0.753
5	0.803	0.938	1.044	0.938
6	0.556	0.720	0.685	0.604
7	0.438	0.607	0.577	0.531
8	0.521	0.582	0.649	0.544
9	1.112	0.994	0.703	0.856
10	0.552	0.556	0.428	0.443
11	0.416	0.423	0.442	0.454
12	0.772	0.737	0.605	0.555

Table 8: Individual VO_2 ($ml{\cdot}kg^{-1}{\cdot}min^{-1}$) values during self regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1	10.140	9.613	14.500	20.607
2	20.207	28.780	20.440	26.600
3	30.713	72.265	26.114	26.350
4	22.087	24.373	19.613	24.688
5	22.120	25.827	28.767	25.667
6	12.527	16.213	15.427	13.609
7	16.787	23.267	22.093	20.359
8	11.787	13.173	14.680	12.313
9	30.147	26.947	19.047	23.194
10	23.675	23.840	18.373	19.000
11	15.033	15.247	15.987	16.395
12	31.400	30.207	24.793	22.731

Table 9: Individual $\%VO_2peak$ values during self regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1	24.71	23.45	35.34	50.27
2	59.94	85.36	60.61	78.91
3	62.04	64.13	56.78	62.39
4	39.35	43.41	34.93	43.95
5	51.39	60.00	66.81	60.00
6	48.25	62.48	59.41	52.43
7	42.66	59.15	56.17	51.76
8	43.30	48.38	53.96	45.24
9	80.44	71.90	50.84	61.92
10	49.99	50.36	38.79	40.13
11	46.60	47.34	49.54	50.86
12	71.10	67.87	55.71	51.06

Table 10: Individual heart rate values during self regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1	87	92	105	121
2	136	161	142	152
3	120	121	117	127
4	127	131	118	131
5	118	126	132	121
6	142	147	141	139
7	121	143	135	135
8	117	126	134	127
9	135	123	108	114
10	125	131	125	126
11	140	143	144	145
12	130	129	119	113

Table 11: Individual %HRpeak values response during self regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1	46.63	48.98	55.94	64.60
2	75.03	89.06	78.67	83.76
3	68.46	69.14	66.86	72.57
4	70.06	72.49	65.19	72.49
5	66.44	71.07	74.58	68.36
6	81.61	84.48	81.03	79.89
7	67.22	79.44	75.00	75.00
8	75.00	80.77	85.90	81.41
9	75.31	68.49	60.45	63.46
10	65.65	68.48	65.34	65.97
11	79.10	80.79	81.36	81.92
12	78.79	78.42	72.00	68.61

Table 12: Individual work rate (W) values during self regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1	5	5	30	55
2	40	70	20	55
3	40	40	35	35
4	30	30	15	30
5	35	45	45	35
6	25	25	15	15
7	5	20	10	10
8	15	20	25	15
9	55	35	20	30
10	20	20	10	10
11	5	5	10	10
12	25	20	10	10

Table 13: Individual %WRpeak values during self-regulated cycling exercise

	Time (min)			
Subject	**0-5**	**5-10**	**10-15**	**15-20**
1	4.35	4.35	26.09	47.83
2	50.00	87.50	25.00	68.75
3	44.44	44.44	38.89	38.89
4	33.33	33.33	16.67	33.33
5	41.18	52.94	52.94	41.18
6	45.45	45.45	27.27	27.27
7	8.33	33.33	16.67	16.67
8	17.65	23.53	29.41	17.65
9	64.71	41.18	23.53	35.29
10	33.33	33.33	16.67	16.67
11	9.09	9.09	18.18	18.18
12	45.45	36.36	18.18	18.18

Appendix II

Definition and Instructions for use of the OMNI RPE scale (78)

Definition: How tired does your body feel during exercise?

Instructions: We would like you to ride on the bike for a little while. Every few minutes it will get harder to pedal the bike. Please use the numbers on this picture to tell us how your body feels when cycling. Please look at the person at the bottom of the hill who is starting to ride a bike (point to the left pictorial). If you feel like this person when you are cycling you will be “not tired at all”. You should point to a 0 (zero). Now look at the person who is barely able to cycle the bike to the top of the hill (point to the right pictorial). If you feel like this person when cycling you will be “very, very tired”. You should point to number 10. If you feel somewhere in between “not tired at all” (0) and “very, very tired” (10), then point to a number between 0 and 10.

We will ask you to point to a number that tells how your whole body feels, then a number that tells how your legs feel and then a number that tells how your chest/breathing feels. Remember there are no right or wrong numbers. Use both the pictures and words to help you select the numbers. Use any of the numbers to tell how you are feeling when cycling the bike.

Appendix III

Physiological and Perceptual Responses During Self-Regulated Exercise in Children with Cystic Fibrosis

Participant Consent Form

1. I confirm that I have read entirely and understand the information sheet pertaining to this study and that all of my questions have been answered satisfactorily.
   (Please tick to indicate yes) ☐

2. I understand that I will be given a copy of the signed consent form and patient information sheet for my records
   (Please tick to indicate yes) ☐

3. I agree to take part in the study.
   (Please tick to indicate yes) ☐

_____________________ ___________ ____________________
Name of participant Date Signature

If the subject is a minor (under 18 years of age):

________________________ ____________ ______________________
Name of parent or guardian Date Signature

To be completed by research team:

____
Date on which subject was supplied with patient information sheet

Dr ___________________ ___________ _____________________
Investigator Date Signature

Information Letter for Parents

Dear Parent/Guardian

My name is Dr, Dubhfeasa Slattery. I am a pediatric consultant in Children's University Hospital, Temple St, Dublin. I am currently undertaking a research study in collaboration with Professor Niall Moyna from Dublin City University (DCU). The title of the study is:

Physiological and Perceptual Responses During Self-Regulated Exercise in Children with Cystic Fibrosis

The OMNI scale is a pictorial-verbal-numerical scale that is used to determine children's feelings of exertion during exercise. The purpose of the study is to determine if the scale is appropriate for use during exercise in children and adolescents with Cystic Fibrosis (CF), and to compare the responses to exercise in children with CF.

As your child is a CF patient in the Children's University Hospital and could participate in this study I would greatly appreciate your support.

Participation in the study will involve 3 separate visits to the Human Performance Laboratory (HPL) in DCU. Each visit will last approximately one and a half hours. During the first visit your child will have his/her height and weight measured, undertake a lung function test and perform an exercise test on a fixed bike to measure his/her fitness. During the second visit your child will undertake 3 separate bouts of cycling at 3 different exercise intensities. Each exercise bout will last 6 minutes and will be separated by approximately 4 min of recovery. During the final visit to DCU your child will cycle for 20 minutes at his/her preferred intensity. Your child will have a mouthpiece in his/her mouth and a clip on his/her nose when pedalling on the bicycle. Your child will also have four electrodes placed on his/her chest to measure heart rate. Following the visits you and your child will be provided with feedback detailing the results of the tests. In addition, you will receive a brief summary of the overall study results.

We hope that the study will provide health professionals with a scale that can be used to prescribe exercise for children and adolescents with CF and that it will also provide important information on the responses to self-selected exercise in children and adolescents with CF.

Your involvement is therefore vital to the success of the study and you and your child's participation would be greatly appreciated. However you are under no obligation to participate in the study. Should you choose to participate anonymity and confidentiality will be guaranteed and your name will not appear in any report or publication arising from the study.

This letter is just to make you aware that there is research taking place among the CF patients in the Children's University Hospital and that a researcher or doctor inviting you and your child to participate in the study may approach you. Permission to undertake the study has been sought through the Hospital Ethics Committee and the relevant hospital staff. Again let me remind you that you are under no obligation to participate in the study and declining to participate will in no way affect the care that your child will receive during their hospital visits.

If you and your child are interested in participating in this study Fionnuala and Eoin will be available to discuss it at the CF clinics or via phone or email.

Contact details

Email: fionnualabritton@gmail.com or eoin.mccormack4@mail.dcu.ie

Phone no: 01 7008470.

Thank you for your time and understanding

Regards

Dr. Dubhfeasa Slattery

____

Physiological and Perceptual Responses During Self-Regulated Exercise in Children with Cystic Fibrosis

Participant Information Sheet

Investigators:

Dr. Dubhfeasa Slattery, The Children's University Hospital, Temple St, Dublin 1

Professor Niall Moyna, School of Health and Human Performance, Dublin City University

1. Invitation

You have been invited to take part in a research study that is being carried out by The Children's University Hospital, Temple St and the School of Health and Human Performance, DCU. The study is being undertaken to measure how your body responds during exercise and to measure how tired you feel when you are exercising.

You do not have to participate in the study if you do not want to. If you decide not to participate in the study, it is important to understand that your care in Children's' University Hospital will not be affected in any way. Please read the following information sheet carefully and feel free to discuss the study with your friends and family. We will contact you to discuss the study further and answer any questions you may have.

2. What is the purpose of this study?

Getting fit and staying fit is important for boys and girls with CF. This study will use a special chart to measure how you feel when you are exercising and it will also measure how your body responds during exercise.

3. Why have I been chosen?

You have been invited to take part in this study because you are a patient in Children's' University Hospital with cystic fibrosis and you are between the age of 8 and 16 years. We plan to have 24 patients with CF in this study.

4. Do I have to take part?

No. Participation in the study is entirely up to you. If you are willing to be contacted about the study, a member of the research team will contact you in the next few weeks to discuss the study further, answer any questions you might have and ask you if you would be willing to take part. Agreeing to be contacted does not mean you have to participate. If you do decide to participate, you will be provided with a consent form to sign. You have the right to stop participating in the study from the study at any time.

5. What does taking part in this study involve?

If you agree to take part in this study we will ask you to visit DCU on 3 separate days. Each visit will last about one and a half hours. During the first visit you will have your height and weight measured, undertake a lung function test and perform an exercise test on an indoor bike to measure your fitness. During the second visit you will exercise four times on the bike. Each cycle will last 6 min and you will have a 4 minute rest after each cycle. During your final visit to DCU you will cycle for 20 minutes at the intensity that you prefer. So that we can measure how your body responds during exercise, you will have a plastic tube in your mouth and a clip on your nose when you are pedalling on the bicycle. You will also have four plastic patches placed on your chest to measure how fast your heart is beating.

6. What are the possible disadvantages and risks of taking part?

There is a possibility that your legs may be tired and a little bit sore and you might feel a bit sick after the maximal exercise test. Occasionally a research subject may be worried or disappointed about some aspect of their results. We will be happy to answer any questions you might have about the various test at any stage of the process.

7. What are the possible benefits of taking part?

You will receive a report detailing the results of your lung tests and fitness evaluations. In addition, you will receive a brief summary of the overall study results.

8. Will my taking part in the study remain confidential?

We would like to assure you that the results of your assessments and tests and any information given will not be shared with anyone other than the doctors and researchers involved in the study. Your identity will remain confidential and your name will not be published. The results of this study will involve looking how the group did as a whole rather than the performance of any one individual.

9. What will happen with the results of the research study?

Dr. Dubhfeasa Slattery will only reveal information about you to yourself and/or legal guardian. It is hoped that the results of this study will help us design appropriate exercise programs for CF patients. We hope to share the information from the study with patients by going through appropriate channels and through publications in special journals. Any information that is shared will discuss group outcomes rather than individual outcomes and your personal details will not be made publicly available.

10. Who has reviewed the study?

The Scientific Committee of the Children's University Hospital has granted scientific approval for this study. Ethical approval for this study has been obtained from the Ethics Committee of the Children's' University Hospital. Prior to participation in the study you will be required to sign a document of informed consent, which will be approved by the Ethics Committee of the Children's' University Hospital.

11. Contact for more information?

Thank you for taking the time to read this information sheet. We would appreciate if you would complete the consent form if you are willing to be contacted by a member of the research team. If you have any questions in the meantime, please do not hesitate to contact the research team at the following telephone numbers.

Please return this section to the research team

I am willing to be contacted by a member of the research team to discuss the research study further. I understand that agreeing to be contacted does not obligate me to participate in the study.

Please tick to indicate yes ☐

Name (print in block capitals) ________________________

Signature: ________________________

Contact number (mobile or daytime number) ________________________

Appendix IV

VISIT 1

Last Name: ________________ **Date:** _________

First Name: _______________ **Subject ID**: _________

Date of Birth: ____/____/____ **Age:** _______ yrs

Contact Details

Contact number (home)______________ Contact number (mobile) ________

Email Address ______________

Informed consent signed Yes ☐ No ☐

Medications:

Nebulized Medication: ____

Inhaled Medication: ____

Oral Medication: ____

Anthropometrics

Height: _________ cm RHR: _____ bpm

Weight: _________ kg BP: ______ mmHg

Skinfolds and % Body Fat Measurement. Tester: ______________

Site	1	2	3	AVG
Tricep				
Subscapular				
Chest				
Midaxillary				
Abdominal				
Suprailiac				
Thigh				
Calf				
Bicep				

% Body Fat : ___________ % Lean Body Mass: _________

Spirometry:

	Absolute	% Predicted
FEV1:	_____	_____
FVC:	_____	_____
MVV	_____	_____

Comments:

Check List

Fasting 4 h	Yes ☐	No ☐
Volume Calibration:	Yes ☐	No ☐
Gas Calibration:	Yes ☐	No ☐
Print calibration:	Yes ☐	No ☐
Informed consent signed	Yes ☐	No ☐
OMNI scale explained	Yes ☐	No ☐
Baseline RPE scale anchor	Yes ☐	No ☐
Peak RPE Anchor	Yes ☐	No ☐

Maximal Exercise Test

Last Name: ________________ **Date:** _________

First Name: _______________ **Subject ID**: _________

Date of Birth: ____/____/____ **Age:** _______ yrs

Height _______ **cm** **RHR** ________ **bpm**

Weight ______ **kg** **BP** ________ **mmHg**

Baseline RPE scale anchor Yes ☐ No ☐

Peak RPE Anchor Yes ☐ No ☐

Stage	Min	Workload (W)	HR (bpm)
Rest			
Warm-up			
Exercise			
1			
2			
3			
4			
5			
6			
7			
8			
9			
10			

Peak Data:

Total time __________ sec

Workload __________ Watts

VO_2 __________ L/min

RER __________

HR __________ bpm

RPE-O __________

Ve __________ L/min

Ventilatory Threshold

HR@VT ___________ bpm

%HR@VT ___________

VO_2@VT ___________ L/min

%VO_2@VT ___________

Workload@VT ___________ Watts

%Workload@VT ___________

Lactate Threshold

HR@LT ___________ bpm

%HR@LT ___________

VO_2@LT ___________ L/min

%VO_2@LT ___________

Workload@LT ___________ Watts

%Workload@LT ___________

Percentage Work Rates

40% VO_2peak ___________

60% VO_2peak ___________

80% VO_2peak ___________

Sub maximal Prescribed Exercise Test

Last Name: ________________ **Date:** _________

First Name: _______________ **Subject ID**: _________

Date of Birth: ____/____/____ **Age:** _______ yrs

Height ______ **cm** **RHR** ________ **bpm**

Weight ______ **kg** **BP** ________ **mmHg**

Contact number (home) ___________________________________

Contact number (mobile) ___________________________________

Email Address ___________________________________

Exercise Bout 1: _____ %VO_2peak

Stage	**Min**	**Workload**	**HR**	**RPE-O**	**RPE-C**	**RPE-L**
Baseline						
Warm-up						
	1					
	2					
	3					
	4					
	5					
	6					
Recovery						
	1					
	2					
	3					
	4					
	5					

Exercise Bout 2: _____ %VO_2 peak

Stage	Min	Workload	Lactate	HR	RPE-O	RPE-C	RPE-L
Baseline							
Warm-up							
	1						
	2						
	3						
	4						
	5						
	6						
Recovery							
	1						
	2						
	3						
	4						
	5						
	6						

Exercise Bout 3: _____ %VO_2peak

Stage	Min	Workload	Lactate	HR	RPE-O	RPE-C	RPE-L
Baseline							
Warm-up							
	1						
	2						
	3						
	4						
	5						
	6						
Recovery							
	1						
	2						
	3						
	4						
	5						
	6						

Sub maximal Self-Selected Exercise Test

Last Name: ________________ **Date:** _________

First Name: _______________ **Subject ID:** _________

Date of Birth: ____/____/____ **Age:** ______yrs

Height ______ **cm** **RHR** ___ **bpm**

Weight ______ **kg** **BP** ___ **mmHg**

Stage	**Time**	**Workload**	**HR**	**RPE-C**	**RPE-L**	**RPE-O**
Baseline						
Warm-up						
1						
2						
3						
4						
5						
6						
7						
8						
9						
10						
11						
12						
13						
14						
15						
16						
17						
18						