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The Physiotherapist's Use of Exercise in the Management of Young People with Cystic Fibrosis

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1. Introduction

The aim of this chapter is to provide an overview of how physiotherapists (physical therapists) can use exercise in their management of young people with cystic fibrosis (CF), assisting the multidisciplinary team to optimise outcomes for this population. While many resources provide management information about the CF health condition, the focus is often on medical management. However, physiotherapists play an integral role in the multidisciplinary team being at the coalface of daily intervention for this population. Consequently, their role - supported by contemporary evidence - must be considered in the holistic approach to management. In this chapter we will briefly discuss the physiotherapists' general management of people with CF, with the focus directed towards the role of exercise in this management regimen.

This chapter explores the benefits of exercise to this population, and considers the factors limiting exercise performance in young people. A review of the previous studies of exercise intervention programs for people with CF in inpatient and outpatient settings will be provided, and issues affecting adherence and clinical applicability will be discussed. A novel exercise program the *Cystic Fibrosis Fitness Challenge* was developed by Mandrusiak & Watter et al. (2009c) and was specifically designed to address the limitations described in the literature. Aspects of this program and its accompanying *FitKit*$_{TM}$ will be presented in this chapter, with an exploration of how the International Classification of Function, Disability and Health – Children and Youth (ICF-CY) (World Health Organization, 2007) was used to frame the selection of performance measures and program elements. Relationships between exercise and other physiotherapy measures commonly reported for this population will also be considered, since applying the ICF framework, we can expect to find relationships between measures within and between domains. For example, impaired cardiorespiratory function may relate to reduced exercise tolerance, which then may be associated with limited activity and restricted participation. However, weak muscles due to decreased activity may also contribute to limited activity, suggesting that management needs to consider impairment, limitations and restrictions holistically rather than as separate issues.

1.1 The ICF framework for describing performance in young people with cystic fibrosis: A basis of design for exercise intervention

This chapter is scoped within the framework of the International Classification of Functioning, Disability and Health – Children and Youth (ICF-CY) (World Health Organization, 2007) which is an extension of the original framework developed in 2001 (World Health Organization, 2001). Because CF is a multifactorial health condition and usually utilizes a team management approach, the ICF-CY provides an excellent framework for a holistic description of assessments, to direct intervention and identify overlaps and gaps in management. In conjunction with family- or client-centred practice, this contemporary approach considers the client with the health condition of CF and their family as the center of management. Of specific interest to physiotherapists in their management of those with CF, relevant *body structures and functions* would include respiratory function, muscle strength and range of motion, *activities* would include motor skills and functional capacity, and *participation* would include activities of daily life at school, home and in leisure pursuits. Further, *contextual factors* include environmental and personal factors (for example, age, gender, inpatient/outpatient status and attitudes towards exercise) and are also considered within this framework. The theoretical underpinnings of the ICF framework state that performance on measures *within* a domain may be related, and further that relationships may exist in measures of performance *between* domains. This implies that changes in respiratory function (*body structures and functions* domain) may be associated with changes in muscle strength (*body structures and functions* domain). Such associations will guide selection of both assessment measures and interventions. As a between-domain example, improvements in respiratory function are expected to relate to improvements in distance walked in the six-minute walk test (*activities* domain) or sports played at school (*participations* domain). Overall, it is proposed that these interactions will provide an important foundation for comprehensive investigation of the performance of young people with CF, and for exploring the impact of contextual factors on their function (Mandrusiak et al., 2009b). Understanding these relationships will provide a strong evidence-base to direct physiotherapy.

2. The focus of the physiotherapist in the management of individuals with cystic fibrosis

The physiotherapist is considered a cornerstone member of the CF multidisciplinary team, and conventionally their management aims to reduce the respiratory impairments related to this health condition by clearing thick, tenacious secretions from the airways (Farbotko et al., 2005). To achieve this, physiotherapy has traditionally focused on airway clearance techniques, and more recently, attention has been given to the integral role of exercise to enhance management and achieve better outcomes across the now extended lifespan of those with CF. In addition, now that we understand the range of impairments that include musculoskeletal issues such as impaired strength (de Meer et al., 1999; de Jong et al., 2001; Hussey et al., 2002) and range of motion (Mandrusiak et al., 2010) , which limit activity and restrict participation in those with CF, physiotherapists are better placed to provide broad based intervention at all levels of the ICF to facilitate optimal outcomes and reduce barriers to participation by these affected children.

The theoretical basis for airway clearance techniques is included elsewhere in this text and will not be considered here. 'Passive' techniques of postural drainage, percussion and vibration, were traditionally the mainstay of respiratory physiotherapy management. These applications were "done to" the individual with CF, and while these techniques continue to play an important role, more active techniques are now available such as the active cycle of breathing technique and positive expiratory pressure devices, as well as inhalation therapy and thoracic mobility exercises (McIlwaine, 2007). This now emphasizes the role of the individual with CF and the family (especially in the case of the young person) in being an active member of the management team and proactively participating in their own treatment, using a "done with the client" or "selected by the client" philosophy. With this trend to encompass more dynamic techniques as a consequence of emerging evidence, physiotherapy now emphasizes the importance of physical exercise as an adjunct to traditional airway clearance techniques. In CF, exercise has other benefits addressing the unique complications which are emerging as longevity improves, such as impairments in posture and bone mineral density. A diagram of the ICF-CY representing the physiotherapists' perspective of possible interventions relevant to the young person with CF is provided below (Figure 1).

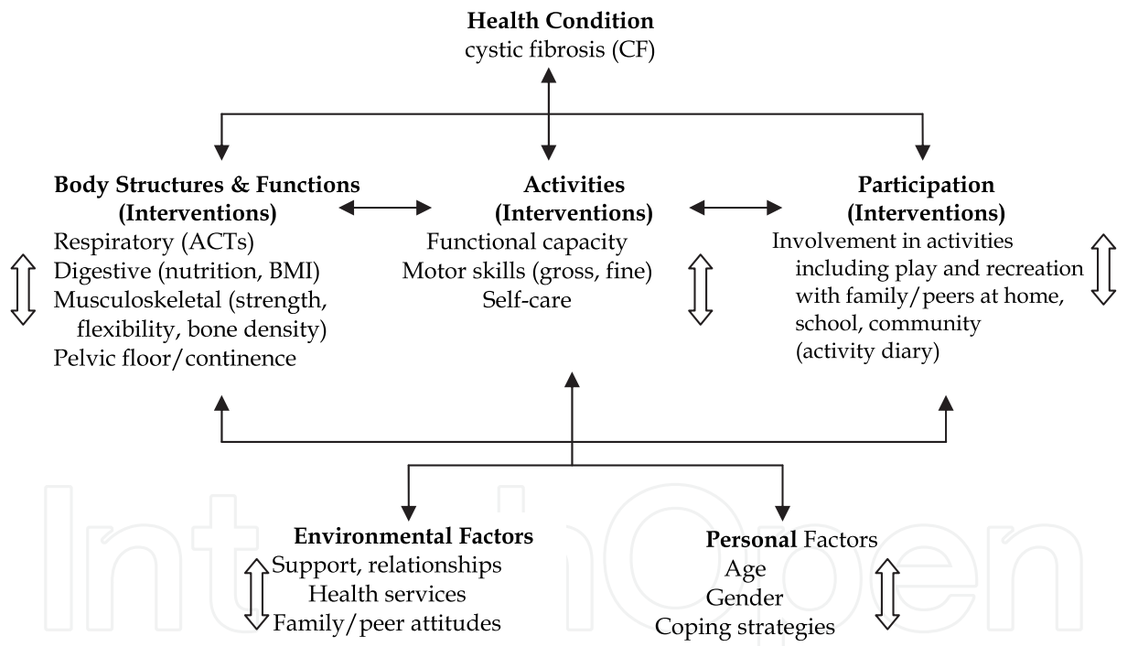


Fig. 1. The ICF-CY (World Health Organisation, 2007): adapted to include possible interventions relevant to the young person with CF.

2.1 Benefits of exercise for people with cystic fibrosis

On the most basic level, exercise is regarded as a natural daily activity for young people, and allows participation in the typical play activities of childhood as well as life-long leisure activities. Ideally, those with CF would be able to partake in exercise with its attendant benefits, and consequently engagement in exercise should be an expectation or goal as well as part of their management. Beyond this basic premise, research and evaluation in the

clinical setting has earned exercise its place in the routine management of people with CF (Dodd & Prasad, 2005). Exercise has long been promoted as an essential element of the care for people with CF because exercise intolerance has always been a trait of disease progression (Orenstein and Higgins, 2005). It is therefore crucial to recognize that participation in regular physical activity is a positive prognostic factor in this population (Nixon et al., 1992). Consequently, promoting exercise for people with CF has become an integral aspect of practice, and its prescription must be based on best evidence.

Cooper (1998 p143) believes the physiological influences of exercise suggest its "*profound and important role as therapy"* for people with CF, and exercise programs show potential to be "*an elegant and natural way to stimulate and/or promote the expression of beneficial genes*". More globally, the psychosocial changes from exercise validate its "*multidimensional impact for people with CF*" (Klijn et al., 2004 p1303). Thus the possibility of potentially modifying the clinical course of the CF health condition by an "*intervention as simple, cheap, safe and enjoyable as exercise remains appealing*" (Barker et al., 2004 p351).

A recent overview of Cochrane systematic reviews performed by Bradley and Moran (2008) summarised the evidence for physical training (exercise) for people with CF. Seven trials using randomised parallel-group design were included, with a total of 231 participants including children and adults. Three studies included children only (Selvadurai et al., 2002; Klijn et al., 2004; Turchetta et al., 2004), and two notable studies included adults and children (Cerny, 1989; Schneiderman-Walker et al., 2000). Overall, these trials showed some evidence of benefits of short- and long-term physical training for people with CF, and this was also the conclusion provided by other reviews in the field (Smidt et al., 2005; Bradley et al., 2006; Shoemaker and Hurt, 2008).

While the effectiveness of exercise intervention programs for this population has traditionally been determined by reduced impairment in body structures and functions (particularly respiratory function) and reduced activity limitations (such as greater six-minute walk distance), the role of exercise for promoting more holistic changes in function is increasingly being recognised. With increased longevity of people with CF, the prevalence of secondary musculoskeletal complications is increasing (Massery, 2005) and must be considered as part of their presentation. This supports the important role of physiotherapy beyond affecting airway clearance, and strengthens the need for an holistic approach which incorporates exercise for prevention or management of secondary musculoskeletal changes (Dodd & Prasad, 2005; Lannefors, 2004; Massery, 2005) to enhance client outcomes. This approach would include addressing range of motion, muscle strength, power and endurance, as well as strong promotion of physical activity. The ICF-CY provides a framework against which such parameters and relationships can be considered.

Further, improving a child's activity and participation are recognized as important goals of exercise intervention for this population, to maintain fitness and thus curtail the cycle of deconditioning associated with this health condition (Stanghelle, 1988; Stevens and Williams, 2007). Importantly, participation in regular physical activity is associated with many holistic benefits, such as improved quality of life and wellbeing (Boas, Danduran, & McColley, 1999; Orenstein, Nixon, Ross, & Kaplan, 1989; Selvadurai et al., 2002). The ICF-CY model facilitates exploration of the impact of exercise intervention on these wider aspects of functioning.

2.2 Providing exercise opportunities across inpatient and outpatient settings: Research supporting advances in practice

Although the performance of exercise is now an integral component of the management of young people with CF, the most effective style of exercise program across inpatient and outpatient settings is yet to be established. During hospitalization, young people with CF are often segregated to minimize cross-infection of respiratory pathogens (Cystic Fibrosis Trust, 2001; Koch et al., 2003), and this presents practical challenges for physiotherapy exercise intervention (Hind et al., 2008). In consequence, it is clear that physiotherapy exercise programs must be adaptable to the limited space at the hospital bedside for performance by individuals in isolation who cannot participate in group exercise sessions or share exercise equipment in the gym. Development of tailored programs which provide a variety of physical activities has been identified as an important aspect to enhance adherence to exercise in this population (Holzer et al., 1984; Blomquist et al., 1986; Stanghelle, 1988; Salh et al., 1989; Abbott et al., 1996; Britto et al., 2000; Schneiderman-Walker et al., 2000; Moorcroft et al., 2004; Turchetta et al., 2004), and such programs must be evidence-based to target issues characteristic of young people with CF.

Developing attractive and efficacious inpatient programs is a challenge in itself, but as highlighted by Moorcroft, Dodd, Morris and Webb (2004) and Dodd and Prasad (2005), transferring exercise programs to the outpatient setting and sustaining them in the long term is also a challenge in the CF population. Physical activity and prescribed exercises are a permanent part of living for those with CF and compliance leading to optimal outcomes is difficult to sustain. Despite these challenges, a range of literature supports the incorporation of outpatient exercise programs in the management of young people with CF. As CF is a lifelong health condition, and the benefits of exercise are well established, strategies to enhance exercise performance across the lifespan are imperative. However, the impact on health resources and clients' distances from established hospital centres make it difficult to facilitate long term supervised hospital-based outpatient exercise programs (Schneiderman-Walker et al., 2000), and sustainable programs based outside of the hospital are needed (Moorcroft et al., 2004).

During outpatient periods, there is support for home-based exercise programs rather than hospital-based programs (Bar-Or, 2000; Bernard & Cohen, 2004; Moorcroft et al., 2004; Schneiderman-Walker et al., 2000; Turchetta et al., 2004). These need to be engaging for the young person with CF, especially if s/he is to continue with these activities as an outpatient once discharged from hospital acute care. Further, it is believed that exercise programs in hospital are not attractive for young people, and that home-based programs are more acceptable (Turchetta et al., 2004). Physical activity should be enjoyable and natural, rather than bear the stigma of therapy, and this may be better achieved in the home environment (Bar-Or, 2000). Further, it is likely that home-based exercise programs save the family time and expense (Bar-Or, 2000) and are more feasible (Bernard and Cohen, 2004). Therefore, research is warranted to establish effective home-based outpatient exercise programs, and facilitate client transition into these programs from the inpatient setting.

2.2.1 Recommendations for exercise programs for young people with cystic fibrosis: Linkages to development of a novel exercise program

A summary of systematic reviews of exercise across a range of populations (Smidt et al., 2005) indicated that targeted and individualized exercise programs were more beneficial than standardized programs. In a Position Statement published by the *Australian Physiotherapy Association* (Taylor et al., 2006) the important skills of the physiotherapist for prescribing exercise are highlighted as three major dimensions: management of disorders of movement, knowledge of exercise regimens and dosages, and clinical reasoning skills to ensure that exercises are optimal for the individual. Specifically for young people with CF, exercise programs should be tailored to individual needs, as there is considerable variability in terms of disease severity, fitness, enthusiasm and preference for types of activities (Webb & Dodd, 1999; Prasad and Cerny, 2002).

To address the aforementioned needs, the *Cystic Fibrosis Fitness Challenge* (*CFFC*) developed by Mandrusiak & Watter et al. (2009c) is a targeted exercise program for use in inpatient and outpatient settings, based on recommendations from the field. Part of this innovative exercise program is a portable exercise tool (*FitKit*™) that is adaptable to limited space environments such as at the hospital bedside in the inpatient setting. The design aspects of this novel program and tools are presented in Section 2.3.

Although specific guidelines are not currently reported, it is recommended that all people with CF should be encouraged to exercise 'several times per week' (Yankaskas et al., 2004), and across the lifespan (Thoracic Society of Australia and New Zealand 2007). Specifically, exercise for *young people* with CF should be viewed as fun as well as therapy and its role should be regarded equally from the young person's point of view as well as a clinical mandate from the multidisciplinary team (Webb & Dodd, 1999). Exercises must be stimulating, age-appropriate and enjoyable, and varied to avoid monotony as well as to avoid overuse injuries (Stanghelle, 1988). Further, prescription of exercise programs should be timely, and appropriate for different settings including within the hospital and the home (Lannefors, 2004). The exercise program must be realistic and allow for the treatment demand and time pressures faced by young people with CF and their families each day, and thus the program must integrate into their lifestyle (Moorcroft et al., 2004). Enthusiasm from the multidisciplinary team members towards exercise and a flexible approach to encouraging physical activities should not be underestimated (Moorcroft et al., 2004).

Educational dialogue is also recommended, as knowledge of the CF health condition and reasons for treatment are associated with increased adherence (Gardner, 2004; Hinton et al., 2002; Prasad & Cerny, 2002). Educational resources must be age-appropriate and thus attractive and colourful (Hinton et al., 2002; Gardner, 2004). These recommendations were integrated into the design of the resources and delivery of the *CFFC* described in Section 2.3.

The ideal elements of exercise programs for this population are presented in the literature, and include: endurance and strength training for the upper and lower limbs; aerobic and anaerobic activities; interval training; weight bearing activities; and flexibility exercises (Webb & Dodd, 2000; Selvadurai et al., 2002; Klijn et al., 2004; Dodd & Prasad, 2005; Bradley et al., 2006; Sahlberg, 2008). As well as addressing the function across these multiple areas, providing a variety of activity types makes it possible to individually tailor the program according to the young person's preferences, thereby improving

exercise participation (Klijn et al., 2004). These elements were considered and incorporated into the *CFFC* program.

As recommended by Rogers, Prasad and Doull (2003), intensity of exercise performance should be derived from results of exercise testing. The "*Clinical Guidelines for the Physiotherapy Management of Cystic Fibrosis: Recommendations of a Working Group*" (Cystic Fibrosis Trust, 2002) recognize that no specific information exists for exercise program intensity and duration for people with CF, and that general recommendations for the 'normal' population are used. These include an intensity of 70-85% of peak heart rate (Orenstein et al., 1981), starting with a duration of exercise classified as 'tolerable' and progressing to 20 to 30 minutes, three- to four-days per week. However, these *Clinical Guidelines* also highlight the differences between children and adults, in relation to exercise ability, indicating that prescriptions for adults may not be appropriate for young people. These differences include: differences in growth, muscle and fat; higher respiratory rate and heart rate; inferior cooling mechanisms; increased energy expenditure and increased reliance on fat metabolism (Cystic Fibrosis Trust, 2002). In young people, strength training must be properly performed, planned and not over-strenuous, as growing bones are sensitive to stress (especially repetitive loading) and the epiphysial plate is susceptible to injury before full growth is complete (Behm et al., 2008). To further minimize these risks it is important to provide a variety of activities to ensure joints are not subjected to repetitive stress (Stanghelle, 1988). Therefore, exercise programs for young people with CF should be specifically designed for this age group, and not just based on those designed for adults.

The health condition of CF affects quality of life in adults mainly due to dyspnea and limitations in exercise capacity which lead to limitations in physical functioning (de Jong et al., 1997). Thus, programs which aim to improve exercise capacity and thus reduce dyspnea may impact positively on quality of life, and it is important to incorporate these strategies into programs for younger people with CF to optimise quality of life across the lifespan.

2.2.2 Fostering adherence to exercise programs in young people with cystic fibrosis

While maintaining adherence to medications and dietary schedules is difficult in the young person with CF and considered elsewhere in this text, our focus as physiotherapists is on fostering adherence to respiratory physiotherapy programs and exercise programs especially as the child with CF enters adolescence and may expect to experience greater autonomy. Overall, people with CF prefer exercise to other components of therapy (Abbott, Dodd, Bilton, & Webb, 1994; Moorcroft et al., 2004; Moorcroft, Dodd, & Webb, 1998), regarding it as a socially acceptable 'normal activity'(Prasad and Cerny, 2002; Orenstein and Higgins, 2005), and an area over which they have control (Abbott et al., 1996). Importantly, adherence to exercise is higher than adherence to respiratory physiotherapy (Schneiderman-Walker et al., 2000). Thus, the integral role of exercise for people with CF is strengthened by its positive perception, which is important to maintaining interest and adherence across the lifespan.

Despite this reported positive perception towards exercise as therapy, the issue of treatment adherence is a growing concern for multidisciplinary CF teams (Kettler et al., 2002). This discrepancy represents a missing link between viewing exercise positively, and adhering to exercise programs. Adherence is a complex and multidimensional issue (Hobbs et al., 2003)

and a detailed review is beyond the scope of this chapter. Instead, key literature in the field of adherence to exercise in young people with CF is presented, to provide strategies for the intervention exercise programs presented here.

Non-adherence to exercise programs may lead to wasted resources, reduced quality of life, missed days at school/work, and higher health care costs (Ireland, 2003; Modi and Quittner, 2003) and thus focusing attention on increasing exercise adherence in this population is critical (Bernard and Cohen, 2004). As children progress into adolescence, the desire to have more control over their lives and a tendency to rebel against authority often result in problems with adherence (Gudas et al., 1991). While strategies to improve adherence in adolescents with CF are integral, it is critical to instill knowledge about exercise during early childhood to enhance adherence into later life stages (Bernard and Cohen, 2004). People tend to look to the short- rather than the long-term benefits of treatment to decide whether to continue (Abbott et al., 1994), so strategies that track progress and improvement in exercise performance, such as log books, pedometry and field tests, may enhance perception of the short-term benefits, and thus enhance adherence.

The multidisciplinary CF team should provide universal support, continued encouragement and education to reinforce the message to the young person and their family that exercise is an important part of treatment (Boas et al., 1999; Dodd & Prasad, 2005; Prasad & Cerny, 2002). Specifically, the role of the physiotherapist for people with CF has been described as that of educator, clinician, researcher and manager (Ireland 2003). The role of educator is particularly important, as empowering patients through education positively affects adherence to treatment and thus health outcomes (Gudas et al., 1991; Hinton et al., 2002; Prasad and Cerny, 2002; Ireland, 2003; Gardner, 2004). Patients are influenced by their own internal belief system; any information or required action needs to make sense and be justifiable to them (Carr et al., 1996). To this end, there is an increasing focus on informing patients and involving them in healthcare decisions (Hinton et al., 2002), and patients should be 'fully involved in any decision-making process during treatment planning' (Chartered Society of Physiotherapy 2000: 8.1). Therefore, using exercise programs that include educational strategies and incorporate the individual in tailoring the exercise program are essential for the physiotherapist to enhance adherence across the lifespan.

An older study by Carr et al. (1996) found that many people with CF (>16 years) did not perceive physiotherapists as having a role in tailoring exercise programs for them. Further, it is reported that exercise prescriptions are often presented in a general manner, without clear specifications of parameters such as frequency and duration (Hobbs et al., 2003). Lannefors (2004) advised that recommending patients to be "physically active" is not enough, and that more active guidance and continuing encouragement is needed. These important findings and the developing evidence base suggest now that physiotherapists need to take a more active involvement in promoting exercise, and in individualizing and monitoring the effects of these programs to strengthen their role in management. Currently, best practice physiotherapy would enhance knowledge of the young person and their family to facilitate ongoing compliance.

The role of parents in influencing their child's participation in exercise is well documented. Lack of parental support towards physical activity may lead to a reduction in regular exercise (Boas et al., 1999), and considering the established relationship between fitness and

prognosis (Nixon et al., 1992; Pianosi et al., 2005), this issue of parent education is of particular relevance. Attitudes toward exercise may be acquired as part of life experience and social support, and with parental encouragement to participate are especially important factors (Baker and Wideman, 2006). Further, parents with an ethos of active personal lifestyle facilitate the young person to be active (Dodd & Prasad, 2005). Education of the family is paramount: a study by Boas et al. (1999) reported that parents of young people with CF perceived fewer benefits of, and greater barriers to exercise than parents of healthy young people, and that less than half of parents in the CF group understood the long term benefits of exercise or knew that exercise performance was related to long term prognosis. However, this education may be improving, as a recent questionnaire provided to 50 young people with CF (8-18 years) and their parents showed they had "substantial exercise knowledge" (Higgins et al., 2007). Overall, there is a need for ongoing comprehensive education of the young person and their family regarding the role of exercise, to achieve optimal outcomes of intervention.

Behavioural strategies to increase adherence to exercise in young people with CF have shown some promise (Bernard & Cohen, 2004; Tuzin et al., 1998). These strategies include self-monitoring, exercising with a partner, behavioural contracting, goal-setting, contingency management, and praise and differential attention. Some of these strategies were incorporated to optimise adherence to the *CFFC* exercise program described in Section 2.3.

As outlined in a review by Dodd and Prasad (2005), there are several perceived barriers to exercise, including unsupportive parental attitudes towards exercise (Boas et al., 1999) and unacceptability of rigid training programs (Gulmans et al., 1999). Further, the daily treatment burden and fatigue associated with the CF health condition make adherence to recommended exercise programs more difficult (Prasad and Cerny, 2002), and this population receives relatively little positive reinforcement for efforts to adhere to treatment (Kettler et al., 2002). This is a driver for physiotherapists when individualizing exercise programs for this population, to ensure integration of physical activity into daily life instead of imposing 'extra' treatment, and to provide appropriate positive reinforcement for participation in exercise. As per the ICF-CY, parents have best impact when facilitatory behaviours towards exercise are displayed.

Specific recommendations for optimising adherence to exercise programs developed for young people with CF are presented in the literature. It is recognised that young people participate more consistently in programs that include a variety of activities, recreational elements and fun (Blomquist et al., 1986), particularly individualized programs which employ activities based on personal preference and perceived competence (Abbott et al., 1996; Britto, Garrett, Konrad, Majure, & Leigh, 2000; Holzer et al., 1984; Moorcroft et al., 2004; Salh et al., 1989; Schneiderman-Walker et al., 2000; Turchetta et al., 2004). In contrast, when the program is regimented, prescribed at an incorrect intensity or not age-appropriate, adherence tends to be poor (Gulmans et al, 1999). Further, programs delivered with specific information about optimal frequency and duration will enhance adherence (Hobbs et al., 2003). An individualised program will facilitate incorporation of regular exercise into their daily life and an already demanding treatment routine (Klijn et al., 2004; Moorcroft et al., 2004). These suggestions to enhance adherence were considered in the development of the exercise program presented later in this chapter.

In summary, addressing adherence issues is paramount to the effectiveness of exercise programs, particularly in a population already burdened by many daily treatments. The multidisciplinary CF team, particularly physiotherapists, must provide support and enthusiasm to promote exercise performance. Educational dialogue is paramount, and should be targeted at the level of the young person with CF and their family. Individualized exercise programs that provide a variety of activities based on the young person's preference and competence, and that allow tracking of personal progress, are more likely to succeed. Overall, this highlights the importance of promoting effective and tailored exercise regimes for young people with CF, and it is vital to include strategies aimed at improving and encouraging adherence.

2.3 A novel approach to facilitating exercise for young people with cystic fibrosis

As outlined above, exercise is a central feature of management in youth with CF due to its positive effects in reducing impairments in cardiorespiratory and musculoskeletal structures and functions, increasing activity and facilitating participation, acknowledging that individual contextual factors may also impact. In response to current clinical challenges and to recommendations in the literature, we developed a novel program (the *Cystic Fibrosis Fitness Challenge* (*CFFC*), and accompanying *FitKit*™) to facilitate performance of exercise integrated into daily life for young people with CF, and design aspects of this program are presented in this section.

The *FitKit*™ (Figure 2) was developed as a portable, tailored resource designed to facilitate exercise performance in a variety of settings - the bedside, gym, inpatient and outpatient - representing an effective tool for physiotherapists working with young people with CF. The portable design of the *FitKit*™ and overall *CFFC* is particularly appropriate in the hospital setting, to support exercise performance where space issues are increasingly common due to segregation of patients for infection control. A variety of exercise elements was included in this program, supported by previous studies in the field, and by findings from earlier studies by Mandrusiak and Watter et al. (2009c) into the presentation of young people with CF in the context of the ICF-CY. A pool of 100 activities presented on colour-coded Activity Cards was developed to address each of the exercise components of aerobic, anaerobic, strength and flexibility. The program facilitator pre-selected the Activity Cards suitable for each participant, and then worked with the participant to select those that they enjoyed, at the same time achieving the therapeutic goals. The *CFFC* involved a 30-60 minute session each weekday over the course of a usual 10-14 day inpatient period, and independently 3-5 days per week at home (outpatient period). Clear guidelines were developed to guide the program facilitators to achieve optimal implementation of the *CFFC.*

A Physical Activity Log (PAL) was designed for participants to record activities performed in each session, during inpatient and outpatient phases of management. Demonstrations for completing the PAL during the inpatient period were provided by the program facilitator, progressing towards self-completion prior to discharge. Information documented in the PAL included exercise type, intensity (for example, heart rate, level of perceived exertion or breathlessness), duration / repetitions, enjoyment level of the activity, limiting factors, pedometry score, and quality and quantity of the sputum expectorated. A sticker system was integrated whereby participants received a gold star for vigorous intensity activities, and a coloured star for moderate intensity activities. This system provided visual feedback

and positive reinforcement regarding the intensity and appropriateness of activities performed, and encouraged participants to monitor and progress performance. Also, the PAL provided visual reminders to the participant to ensure adequate hydration during each session. Educational strategies were incorporated into the resources in the *FitKit*™ to increase awareness within this population about the role of exercise (Gudas et al., 1991; Hinton et al., 2002; Prasad and Cerny, 2002; Ireland, 2003; Gardner, 2004) during inpatient and outpatient periods, with emphasis on monitoring exercise intensity (heart rate, and to be aware of exertion and breathlessness levels) to ensure inclusion of activities of vigorous intensity. Overall, the *FitKit*™ is feasible, utilizing inexpensive and readily available resources and equipment (Figure 2), which has significant clinical implications (Orenstein & Higgins, 2005).



Fig. 2. The portable *FitKit*™ used in the *Cystic Fibrosis Fitness Challenge* program

The *CFFC* program provides an evidence–based novel approach to facilitating performance of physiotherapy exercise programs, across both inpatient and outpatient settings. A randomised controlled trial (Mandrusiak & Watter et al., 2009c) showed the effectiveness of this program during inpatient (10-14 days) and outpatient (8-12 weeks) phases of management in a group of young people with CF 7-17 years of age (n=31).

To summarize the recommendations that have been integrated into the design and delivery of this program, the *CFFC* provided a variety of age-appropriate activities that can be tailored to individual preferences across inpatient and outpatient (home) settings,

incorporating education, communication and behavior modification strategies. The overall aim is to enhance integration of physical activity into daily life, and thus achieve long term adherence and optimal outcomes. Individualized exercise programs that provide a variety of activities based on the young person's preference and competence, and that allow tracking of personal progress, are more likely to succeed, and these elements were embedded into the design of the *CFFC* and *FitKit*™.

2.4 Overview of the role of exercise testing for young people with cystic fibrosis: What do physiotherapists measure?

A holistic range of measures which map to all of the ICF domains described above are provided by various professionals in the team managing those with CF, including measures of respiratory function, diet, medication, musculoskeletal function, as well as activity and quality of life. The physiotherapist is not necessarily involved in collecting them all, but is most likely to collect data about body structures and functions such as respiratory function tests and musculoskeletal measures, activity and functional exercise capacity. The focus of this section is on exercise testing.

Exercise testing is the global assessment of the response to exercise, and is an important parameter that is inadequately reflected by resting respiratory function tests (Baraldi and Carraro, 2006). It is an important outcome measure (Stevens and Williams, 2007) which may be more sensitive to disease progression and survival than respiratory function tests (McIlwaine, 2007), and specifically, aerobic fitness is a reliable indicator of disease status and prognosis (Nixon et al., 1992). For young people with CF, measures of activity performance and exercise testing can provide valuable information about the impact of the disease, functional limitations and trends over time (Rogers et al., 2003; Barker et al., 2004), and it is employed as an outcome variable in some intervention studies (Orenstein and Higgins, 2005). Further, Rogers et al. (2003) suggest that regular exercise testing accentuates the value of exercise to young people with CF and their families, which may encourage active lifestyles.

The guidelines for the *Association of Chartered Physiotherapists in Cystic Fibrosis* (in Rogers et al., 2003) recommend that all young people with CF should have annual exercise testing. Measurements of exercise capacity should provide the foundation for any prescription and adaptation of exercise programs (Rogers et al., 2003). Currently, exercise capacity is assessed using a range of laboratory- and clinically-based tools including cycle ergometry, walk tests, step tests and shuttle tests. Debate remains as to the most effective way of determining exercise capacity (Rogers et al., 2003), and some of the most commonly used measures are discussed below.

2.4.1 Maximal and submaximal tests of capacity

Maximum oxygen consumption (VO_2max) is the best index of aerobic capacity, and is significantly correlated with subsequent survival in people with CF (Pianosi et al., 2005). VO_2max is the "gold standard" measure of cardiorespiratory fitness, and maximal tests are performed in a laboratory using cycle ergometry or treadmills (Bruce, 1971). However, a number of factors limit the application of maximal tests to young people. Firstly, most daily activities are not performed at maximal levels and instead are at moderate intensities

interspersed with short bursts of high intensity activities (Bailey et al., 1995), thus using maximal tests may not provide a realistic simulation of a young person's physical capacity (Chetta et al., 2001; Rogers et al., 2003; Solway, Brooks, Lacasse, & Thomas, 2001). Secondly, maximal tests are tiring and the physiological stress of testing, as well as the possible safety risks and expense to the patient, may outweigh the information gained (Nixon et al., 1996), and many young people with CF are reluctant to perform them (Rogers et al., 2003). Thirdly, data from maximal exercise tests may not be reproducible (Stevens and Williams, 2007). Finally, these tests are usually performed by specialist respiratory personnel and require sophisticated laboratory equipment that may need to be modified for young people and may not be available in all institutions (Orenstein, 1998; McIlwaine, 2007). In view of these issues, maximal exercise testing is currently outside the scope of routine clinical physiotherapy practice (Thoracic Society of Australia and New Zealand, 2007), and is not discussed further here.

According to Braggion (1989), submaximal field tests are better tools than maximal cycle ergometry or treadmill tests to evaluate a range of parameters including cardiorespiratory adaptations to exercise, motor aspects of performance (including agility, muscle strength and range of motion), and motivation to perform. Further, while field-based exercise tests do not always determine a person's maximal exercise response, they can give valuable clinical information on factors that limit activity performance on a day-to-day basis (Noonan and Dean, 2000; Narang et al., 2003; Rogers et al., 2003). Field tests are attractive to clinicians and researchers as they provide easy to administer and inexpensive forms of exercise assessment using typical activities of daily living such as walking (Orenstein, 1998). These can be undertaken outside of formal testing laboratories which may promote a less stressful environment for the young person (Cox et al., 2006) and some are useful in the research context where portable tools can provide follow-up information without the need for repeated visits to hospital facilities. Further, there is some evidence that young people with CF prefer field tests to formal exercise tests (Selvadurai et al., 2003). However, it is important to recognize that some information which is detected by more complex exercise tests may be missed by simple field tests (Narang et al., 2003).

In summary, young people rarely engage in sustained, heavy exercise, suggesting that traditional maximal exercise tests may not represent their patterns of daily physical activity (Cooper, 1995). Submaximal tests may better simulate childhood activities and thus provide insight into their functional capacity. These performance measures must be simple and convenient to use in order to be applicable to a variety of settings, such as the clinic, the hospital bedside and in the field (Narang et al., 2003), and such tests include walk tests, step tests and jump tests.

2.4.1.1 Walk tests

A variety of walk tests exist, but the six-minute walk test (6MWT) (Butland et al., 1982) is endorsed as the safest and easiest to administer, and it is better tolerated and better reflects activities of daily living than other walk tests such as the shuttle walk test (American Thoracic Society, 2002). It is an important clinical assessment tool, since it provides a composite assessment of respiratory, cardiac and metabolic systems during exercise (Li et al., 2005). It is self-paced and assesses the submaximal level of functional capacity, where the participant chooses their own level of intensity (American Thoracic Society, 2002). This self-

paced nature may more closely reflect functional performance than externally paced exercise tests such as shuttle tests (Solway et al., 2001). Further, the use of a standard time (six-minutes) rather than a predetermined distance provides a better measure of endurance (McGavin et al., 1976).

Butland et al. (1982) revised the original 12 Minute Walk Test, to better accommodate patients with respiratory disease for whom walking for 12 minutes is too exhausting. The resulting 6MWT was found to perform as well as the 12 minute walk test, and is now a widely used measure for young people with CF, as it is reproducible (Gulmans et al., 1996; Balfour-Lynn et al., 1998; Cunha et al., 2006; Mandrusiak et al., 2009a), valid (Gulmans et al., 1996) and easy to perform in young people with CF (Cunha et al., 2006). It has been studied in a range of CF cohorts including inpatients (Mandrusiak et al., 2009a) and outpatients (Butland et al., 1982; Cunha et al., 2006; Gulmans et al., 1996).

The primary measurement is distance walked in six-minutes (6MWD), but data can also be collected about oxygen saturation (SpO_2), heart rate (HR) and breathlessness (Enright, 2003). Also, 'work' can be calculated as distance walked (m) x body weight (kg) (Chuang et al., 2001) and is recommended instead of distance as it more accurately indicates true performance (Cunha et al., 2006).

As summarised by Noonan and Dean (2000), the 6MWT can be employed as a one-time measure of functional capacity, or to measure change in functional capacity over time or in response to intervention. In people with CF, the 6MWT has been validated, being compared with cycle ergometry (Gulmans et al., 1996) and the 3min step test (Balfour-Lynn et al., 1998). A significant improvement in 6MWD was found in young people with CF at completion of hospital treatment for acute respiratory infection (Upton et al., 1988). Although no minimal clinically important difference (MCID) (Guyatt et al., 2002) for 6MWD has been established for the CF population, in adults with chronic obstructive pulmonary disease an improvement of 70 meters walked after an intervention is necessary to be 95% confident that the improvement was significant (Redelmeier et al., 1997).

Li et al. (2005) found a significant correlation between the 6MWD and VO_2max on the treadmill in typical children. It was also reported that in people with CF, 6MWD correlated with VO_2max, physical work capacity and the minimum arterial oxygen saturation (SaO_2) (Nixon et al., 1996), as well as with forced expiratory volume in one second (FEV_1) (Geiger et al., 2007).

Geiger et al. (2007) presented a modified 6MWT in which the participant pushed a measuring wheel, to establish reference values of healthy young people (n=528; 3-17 years). Li et al. (2005) presented height-specific reference values from a cohort of healthy (Chinese) children (n=1445; 7-16 years). 6MWD was related to height in some studies (Nixon et al., 1996; Cunha et al., 2006) but was not in other studies (Bradley et al., 1999), and correlated with weight by Gulmans et al. (1996). Although complete data has not been developed and consensus is not reached on all issues, it is important to consider factors relating to growth when comparing repeated results for young people with CF over time, and also comparing young people with CF to normative values.

A limitation of the 6MWT is that it requires an uninterrupted corridor of at least 30 meters, so is not adaptable to all settings. Consequently, this test may not be suitable for

performance at the hospital bedside or in the home, where possible space limitations may not ensure standardized administration. Hence, tests that correlate with the 6MWT but require less space may present attractive alternatives. However, where applicable, the 6MWT appears to be the preferred tool for assessing exercise capacity in young people with CF.

2.4.1.2 Shuttle tests

The modified shuttle walk test as described by Selvadurai et al. (2003) is a symptom-limited exercise test, in which the participant moves from end to end of a 10m course in time with the 'beeps' from a pre-recorded tape. This test is valid in young people with CF (Cox et al., 2006), sensitive to change after hospitalization (Bradley et al., 1999, 2000) and correlates with VO_2max (Rogers et al., 2003). It is a natural activity and easy to administer, but does require 10 meters of uninterrupted space, and its practicality in the clinic setting has been questioned (Balfour-Lynn et al., 1998). Thus, motor tasks which allow more efficient delivery, and are adaptable to a range of clinical environments such as the hospital bedside or the outpatient clinic cubicle, may provide a suitable alternative.

2.4.1.3 Step tests

The three-minute step test is an externally paced, simple and portable test which is independent of effort and validated for use in children over six-years of age (Balfour-Lynn et al., 1998). It detects improvement in exercise capacity following hospitalization (Pike et al., 2001), but its use may be limited in those with well preserved lung function and fitness levels due to a potential ceiling effect (Selvadurai et al., 2003). It is also clear that actual workload will vary according to step height and weight and height of the participant (Selvadurai et al., 2003).

2.4.1.4 Jump tests

Mandrusiak et al. (2009a) established test-retest reliability of two jump tests (Astride Jumps and Forwards-Backwards Jumps) for young people with CF in the inpatient setting, using motor measures (number of jumps, time to fatigue) and physiological measures (heart rate, oxygen saturation via pulse oximetry, Borg Rating of Perceived Breathlessness (Burdon et al., 1982), and 15 Count Breathlessness Score (Prasad et al., 2000)). These jump tests are reflective of the natural activity pattern of typical young people, which is characterized by short bursts of high intensity activity (Bailey et al., 1995), and are particularly appropriate for the clinical setting as they are portable and easy to administer in limited space environments such as at the hospital bedside or outpatient clinic room. The score is the number of jumps performed before fatigue, and this score has been shown to improve significantly after hospitalization in 33 young people with CF (7-17 years) (Wilson et al., 2005). Work is being conducted to establish references for Australian children for such simple activities used in daily life.

In summary, measures of physical performance and exercise capacity are integral to the management of young people with CF. Tests for this population should be non-invasive, simple and quick to administer, inexpensive and applicable to a variety of settings (Cooper, 1998), and reflective of typical activities of childhood. In our experience, children tolerated and enjoyed the 6MWT and jump tests, and these are user-friendly field tests.

3. Conclusion

This chapter concerns the integral role of exercise in the management of young people with CF, providing a contemporary overview of current research and practice as well as the existing limitations and gaps to direct future research. The innovative *Cystic Fibrosis Fitness Challenge* and *FitKit*$_{TM}$ developed by the chapter authors presented a working example of how the ICF-CY framework can direct selection of performance measures and guide development of a program, as well as assessment of effectiveness of the intervention. This contemporary information supports the evidence base for the role of exercise in the management of those with CF and is an essential aspect of the physiotherapy role within the multidisciplinary team.

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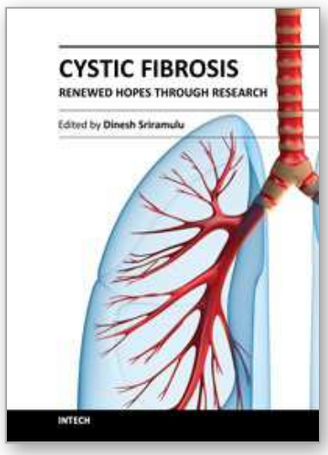
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Living healthy is all one wants, but the genetics behind creation of every human is different. As a curse or human agony, some are born with congenital defects in their menu of the genome. Just one has to live with that! The complexity of cystic fibrosis condition, which is rather a slow-killer, affects various organ systems of the human body complicating further with secondary infections. That's what makes the disease so puzzling for which scientists around the world are trying to understand better and to find a cure. Though they narrowed down to a single target gene, the tentacles of the disease reach many unknown corners of the human body. Decades of scientific research in the field of chronic illnesses like this one surely increased the level of life expectancy. This book is the compilation of interesting chapters contributed by eminent interdisciplinary scientists around the world trying to make the life of cystic fibrosis patients better.

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