Exercise and Habitual Physical Activity for People With Cystic Fibrosis: Expert Consensus, Evidence-Based Guide for Advising Patients

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Exercise and physical activity have been recommended since the 1970s as important aspects of care for people with cystic fibrosis (CF). Benefits of an active lifestyle for these patients include improved pulmonary function, functional capacity, and quality of life and also decreased mortality. However, there are not currently any guidelines available to CF care centers regarding the type, amount or frequency of exercise, or habitual physical activity that should be performed to achieve these benefits. In addition, there is no guidance available about adapting physical activity and exercise for different ages or comorbidities common to CF. This document presents evidence-based recommendations from an international group of experts in the area of exercise for CF to guide practitioners toward the goal of regular, appropriate habitual physical activity, exercise and sports for all patients with CF. As lifespan for people with CF increases, it is important that regular physical activity is incorporated as part of daily care for a lifetime. **(Cardiopulm Phys Ther J. 2015;26:85–98)** *Key Words: cystic fibrosis, exercise, physical activity, prescription, precautions*

INTRODUCTION

Cystic fibrosis (CF) is a genetic disease affecting approximately 30,000 Americans and 70,000 people worldwide.[1] The disease is caused by a malfunction in the cystic fibrosis transmembrane regulator (CFTR) gene, which leads to the formation of abnormally thick, sticky mucus in the lungs, digestive tract, and reproductive tract.[1] People with CF typically develop respiratory tract infections and progressive obstructive lung disease and also malnutrition. Currently, the disease is ultimately fatal, typically because of respiratory or cardiac failure, at a mean age near 40 in the United States[1] and over age 50 in Canada.[2] However, as more is known about the genetics and diverse phenotypic presentations of CF, many patients are living well into adulthood.

Habitual physical activity and exercise have been shown in multiple studies to positively impact lung function,

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decrease pulmonary exacerbations and hospitalizations, and improve physical function, endurance, energy level, and quality of life for both children and adults with the disease. It is currently considered part of standard CF care to encourage a physically active lifestyle.[3–9] Indeed, many patients report regular physical activity and exercise to be critical in helping them participate in a normal life. Recently, a large study of over 1000 people with CF found that self-report of participation in exercise was associated with better longitudinal nutritional and pulmonary outcomes in adults with the disease, compared with those who were not exercising.[10]

However, to date, little guidance has been provided regarding the appropriate types and dosage of habitual physical activity or exercise necessary to achieve positive outcomes. Also, little guidance is available to modify physical activity for people with CF with complications. Thus, the CF care team, particularly the team physical therapist (physiotherapist), is charged with helping patients become physically active and maintain physical activity with little guidance.

This document is intended to provide the CF care team, particularly exercise professionals who are responsible for the physical activity component, with evidence-based guidelines to provide the framework for patient counseling in a physically active lifestyle. The document grew from initial meetings of the Cystic Fibrosis Exercise Working Group of the European Cystic Fibrosis Society and the Exercise Special Interest Group meetings at the North American Cystic Fibrosis Conference, respectively. This group initially met in 2008 and has been working since that time to develop evidence-based clinical practice guidelines in the area of exercise and habitual physical activity. The group, consisting of approximately 50 physicians, physical therapists, and exercise scientists, has developed position statements on assessment of habitual physical activity in CF[106] and also exercise testing for people with CF.[107]

The current document is intended as a companion to these previous documents, which will allow exercise professionals and CF caregivers to translate the results of exercise testing and habitual physical activity assessment into practical recommendations for patients across the lifespan and disease spectrum of CF. The format of the document arose from a series of clinical questions posed by the group, based on inquiries from patients, families, and CF care team members regarding exercise guidelines. The primary writing group, identified for their clinical and research expertise in the area of exercise and CF, was nominated and undertook the initial draft of the recommendations. The document was then vetted through the larger group, using a modified Delphi process to solicit feedback.

Key Definitions and Format for this Document

To guide exercise professionals toward appropriate exercise and physical activity prescription for people with CF, we will use the following definitions throughout the document:

- Habitual physical activity: bodily movement produced regularly by the contraction of skeletal muscles that result in a substantial increase over resting energy expenditure[11]
- Exercise: planned, structured, and repetitive bodily movement performed to improve or maintain one or more components of physical fitness[11]
- Sport(s): an activity involving physical exertion and skill in which an individual or team competes against another
- Young child: ages 1–6
- Child: ages 7–12 (prepubescent)
- Adolescent: ages 13–18
- Adult: ages 19 and greater.

Effect of CF on Exercise Responses

A normal exercise response depends on a complex interaction of 3 major systems: pulmonary, metabolic, and cardiovascular. In CF, impaired lung function and obstructive lung disease alter the ventilatory responses to an exercise challenge. Currently, it is unknown precisely what level of lung damage limits exercise, but expert opinion, based on evidence,[12,13] suggests that if forced expiratory volume in 1-second (FEV_1) is less than 50% of predicted values, the patient may be limited in his or her aerobic exercise capacity by ventilatory factors. Anaerobic exercise, which uses either short burst, high-intensity activity, or other types of exercise that use smaller amounts of muscle mass (eg, resistance training), is likely not limited by ventilatory factors.

The digestive system impairments of CF lead to impaired nutritional status and low body mass. In particular, people with CF often have less skeletal muscle mass than peers of normal weight. In addition, the CFTR protein has been identified in skeletal muscle, suggesting that the disease itself could impact muscular function.[14] A major component of a normal exercise response depends on the ability of skeletal muscle to use oxygen and other nutrients to produce muscular work. Given that muscle size is directly related to force output, both lack of lean muscle mass and impaired metabolic function has been seen in CF and can impair exercise response. Again, there is no definitive value below which these muscle impairments would impair exercise capacity, but the exercise professional must be aware that severely low lean body mass would make prolonged or whole-body exercise quite difficult. In addition, it is important that the exercise professional works closely with the CF dietitian to optimize not only total caloric intake but also the quality of calories consumed to compensate for the demands of exercise. The muscles of ventilation also contribute to the exercise response. In CF, there is suggestion that significant obstruction and significant malnutrition may impact both strength and endurance of these muscles and may contribute to impaired ventilatory capacity with exercise.[15] It is important to identify deficits in ventilatory muscle performance, as these may be at least partially alleviated with targeted ventilatory muscle training.

Cardiovascular function is the third major component that contributes to exercise capacity. In CF, little impact of cardiac issues is generally seen, except in the case of heart

failure secondary to severe lung disease (ie, cor pulmonale). However, it is common to see elevated heart rates at rest, particularly in very lean or sedentary patients. A high resting heart rate limits the reserve to increase cardiac output with exercise and may lead to premature cessation of higher intensity activities. Peripheral vascular dilation has been found to be impaired in one small study of children with CF[16] but not in a small study of adults with CF.[17] These findings suggest that blood flow to exercising muscles may be abnormal, at least in some patients.

Other factors that may contribute to poor exercise capacity in this population include medications and low levels of physical activity. Many patients receive systemic corticosteroids at some point in the management of their disease, and these medications are well known to impair muscle function.

In the guidelines that follow, we format the recommendations as responses to clinical questions (eg, how much aerobic exercise should be performed by a child with CF?) to help exercise professionals quickly identify the recommendation most appropriate for an individual patient being counseled. Each section lists a series of clinical questions, followed by the recommendation and supporting evidence and/or rationale for the recommendations. A summary of the recommendations by age group is found in Table 1.

Recommendations Regarding Habitual Physical Activity for People With CF

Clinical Question. How much habitual physical activity should be performed by a young child with CF?

Recommendation. Young children with CF should perform regular, developmentally appropriate physical activity at least 60 minutes per day.

Evidence for Recommendation. This guideline is based on recommendations for healthy children ages 5–17.[18] There is little evidence specific to CF; however, given that most young children have little disease-related limitations to their physical activity, the experts believe the guideline is appropriate. In fact, physiotherapy treatment for asymptomatic infants and toddlers with CF is increasingly focused on structured daily physical activity.[6] Specific considerations for the young child include making the physical activity developmentally appropriate, such as including time spent in the prone position for infants to develop trunk extensor muscles and posture. Toddlers and preschool aged children should be performing a wide variety of play activities, including high-intensity, short-burst activities, because this best suits their metabolic capacity. Physical activity should be fun, and parents and siblings should be encouraged to participate along with the child. Having a child or sibling with CF can be quite stressful, and physical activities performed together can provide stress relief and allow a sense of normality. Parents should be encouraged to allow their child to experience a wide variety of physical activities, exercise, and sports to expose them to options they may wish to pursue later in childhood and adolescence. The goal at this age is to make habitual physical activity a family activity, as young children are heavily influenced by the family culture. Also, it is important to introduce physical activity as part of treatment of CF early (eg, at diagnosis) because the burden of CF care will often increase with age, and it can be difficult to begin later in life.

Clinical Question. How much habitual physical activity should be performed by a child with CF?

Recommendation. Children with CF should engage in at least 60 minutes of moderate-to-vigorous physical activity daily.

Evidence for Recommendation. Children with CF have been found to engage in lower levels and intensities of habitual physical activity than their peers,[19] particularly if they had moderate or severe lung disease[20] or impaired nutritional status.[21] Although there has been little study in this age group specific to CF, studies have found weak or no correlation of habitual physical activity level to peak aerobic capacity,[22] suggesting a more complex interaction between these 2 variables. Children with CF were found to value physical activity at the same level as healthy children after a 2-week training program,[23] suggesting that habitual physical activity can be viewed positively. Recently, a large study in CF found that self-reported exercise participation rates peaked at age 10,[10] suggesting that childhood is a vital time to develop physical activity habits. The guideline, therefore, is based on recommendations for healthy children of the same age, as well as results of assessment recommendations for CF.[18,106] The physical activity for a child with CF should be at least intense enough to make the child feel like he/she is breathing somewhat hard but can still carry on a conversation. This intensity is important, not only for the general health benefits but also for airway clearance. More intense activities including strengthening or interval training may induce greater gains in muscular adaptations and physical fitness. The types of physical activities should be those that the child enjoys and need not be continuous in nature. Running, jumping, and ball games such as basketball or volleyball also involve significant impact forces that may help build or maintain bone density at this age. Because bone mineral density (BMD) loss is a significant issue in later adulthood for people with CF, it is recommended to maximize the so-called "bone bank" at this age and in adolescence. Resistance training is also recommended for this age group as one option for regular muscle strengthening,[18,24] provided that children are properly supervised and technique is emphasized over amount of weight lifted.

Clinical Question. How much habitual physical activity should be performed by an adolescent with CF?

Recommendation. Adolescents with CF should perform at least 60 minutes of moderate-to-vigorous physical activity daily.

TABLE 1

Summary of Recommendations for Physical Activity by Age Group

Type of Activity	Young Child (1–6 yrs)	Child (7–12 yrs)	Adolescent (13–19 yrs)	Adult (>19 yrs)
Habitual physical activity	60 min daily in developmentally appropriate activities	60 min daily in a variety of activities enjoyed, preferably as a family activity	60 min daily in a variety of activities enjoyed, especially with family or friends	150 min per week or more (preferably 300 min) in a variety of activities of choice
Aerobic exercise	No formal program recommended but should perform full-body activities that increase ventilation and heart rate	30–60 min daily of moderate-to-vigorous exercise (at least 70% of maximal heart rate) especially if using for airway clearance (must also perform coughing/huffing to clear secretions)	30–60 min daily of moderate-to-vigorous exercise (at least 70% of maximal heart rate) especially if using for airway clearance (must also perform coughing/huffing to clear secretions)	30–60 min daily of moderate-to-vigorous exercise (at least 70% of maximal heart rate) especially if using for airway clearance (must also perform coughing/huffing to clear secretions)
Resistance/strengthening exercise	No formal program recommended but should perform activities using body weight to develop strength (eg, calisthenics)	All activities that use body weight to strengthen muscles and bones (eg, calisthenics) most days and if interested, begin formal weight training program under good supervision, learning technique first (twice weekly)	Formal resistance training 2–3 times per week per muscle group, incorporate limb and trunk muscles, 1–3 sets of 8–12 repetitions at 70%–85% of maximal load (1-repetition maximum)	Formal resistance training 2–3 times per week per muscle group, incorporate limb and trunk muscles, 1–3 sets of 8–12 repetitions at 70%–85% of maximal load (1-repetition maximum)
Other comments	Encourage normal motor development, including agility and balance/coordination	Encourage normal motor development, including agility and balance/coordination	Encourage muscle activities to help prevent or minimize adverse postural changes	Adapt for disease-related complications such as CF-related diabetes or low bone density as indicated

CF, cystic fibrosis.

Evidence for Recommendation. Adolescence is a challenging and exciting time for parents and emerging young adults. For adolescents with CF, puberty is often delayed and they may feel embarrassed by physically lagging behind their peers. In addition, physical activity levels are lower in this age group,[25] particularly in girls with CF. Low levels of physical activity in girls aged 7–17 with CF were associated with steeper rates of decline in FEV_1 over a 2-year study period,[26] whereas higher levels of habitual physical activity were associated with slower rate of decline for boys and girls together.[27] The unique concerns of CF, such as coughing, expectorating, flatulence, or leakage of urine (which often occur during physical activity) and body image concerns may make adolescents with CF even more likely to decrease physical activity. Furthermore, adolescents' behavior is increasingly influenced by peers and family members, and if their peers are sedentary, there may be a lack of desire for the adolescent with CF to be physically active. For some adolescents, however, this may be a time for participating in team sports or physically active recreation that can help reinforce regular physical activity (eg, outdoor adventure activities). For adolescents who are smaller or have less muscular development than their peers, however, they may find that they are no longer able to keep up the emerging demands of competitive team sports. Families need to continue to encourage outlets for vigorous physical activity, as a routine part of CF care, just like taking prescribed medications and performing regular airway clearance. With counseling on physical activity choices that best fit the adolescent's interests and abilities, long-term adherence and adoption of a physically active lifestyle have been demonstrated.[28] Particularly for girls, including adolescents, participation in habitual physical activity has been shown to slow the rate of decline in lung function, and lower levels of habitual physical activity may partially explain the poorer survival of females with CF.[26]

Adherence to all care guidelines can be challenging at this period, and the exercise professional needs to consider ways for the adolescent to have choice in types of activities, timing, etc., to maximize the chance of adherence to the

guideline. At least one study found that free-choice physical activity participation was correlated with maximal aerobic capacity,[29] suggesting that mode of physical activity may not be as important as choosing activities in which the adolescent will agree to engage. Encouraging patients to view (appropriate) websites and online chat groups targeted to teens with CF may help them see the value of physical activity. Many high-profile people with CF who can be seen or reached through these websites have indicated the value of exercise and physical activity in maintaining their physical and emotional health through this, sometimes difficult time of life.

It is important that exercise professionals speak directly to the adolescent, ideally without the parents' presence, to establish rapport and openness to questions and concerns the adolescent may have about exercise, particularly "fad" or trendy programs, and to talk about the dangers of performance enhancing substances that may be offered to gain weight or muscle mass. By pointing the adolescent to peers with CF (such as online groups), the exercise professional must be aware of both the positive and negative influences to which the adolescent may be exposed and talk frankly with the patient about these issues. The goal in this period is to maintain physical activity as a desirable and positive response to having a chronic illness (eg, this is something the adolescent can control) and help lay the foundation for lifelong physical activity habits.

Clinical Question. How much habitual physical activity should be performed by an adult with CF?

Recommendation. Adults with CF should perform at least 150 minutes per week, and preferably 300 minutes per week, of moderate-to-vigorous physical activity.

Evidence for Recommendation. This recommendation is based on the most recent physical activity guidelines[18] and expert recommendations for adults with CF. Data specific to adults with CF have shown that they have lower levels of habitual physical activity than their peers, particularly in more vigorous levels.[30–32] It has been suggested that declines in habitual physical activity in adolescence carry over into adulthood.[32]

There are many examples in the lay literature and Internet sources (eg, Boomer Esiason Foundation) of adults who attribute regular, and quite vigorous, physical activity and sports performed throughout their lives as the key to managing life with CF. All adults with CF must learn to manage the demands of their disease and its care. Many find that exercise and physical activity are, indeed, "treatment for the disease"—important for airway clearance, appetite and weight management, pain management, and stress relief—and put exercise and physical activity foremost in their daily CF care routine.

As adulthood will encompass a wide array of disease-related issues, physical activity at this stage of life must be enjoyable and adaptable to changing health status and must fit into a busy life (for practical tips, see www.cff.org/adults/dailylife/exercise). If adults experience the benefits of being physically fit, this can be a critically important motivator for positive self-care. Studies have shown that adolescents and young adults who are regularly physically active maintain their lung function capacity longer than those who are sedentary and have lower rates of hospitalization than those who are more sedentary.[26,33] Discussing these proven benefits with a patient who is currently sedentary can be an important motivator for change. It is important to guide patients in a reasonable plan to reach the ultimate recommendation above, targeting the counseling approach to their specific facilitators, barriers, and attitudes toward physical activity and exercise.[34,106]

Considerations for Habitual Physical Activity Across the Lifespan

Cystic fibrosis is an ever-changing disease and to develop and maintain a habit of physical activity, the exercise professional must be able to help the patient adapt. The intensity and type of physical activity must be adapted for the patient who is experiencing a pulmonary exacerbation. Progressive lung disease may mean that a preferred sport from childhood may not be possible later in life. The exercise professional needs to be able to analyze different types of physical activity for their muscular, ventilatory, and cardiac demands to recommend new options. Environment (eg, weather, seasons, and altitude) will influence the demands of physical activity on the body. The exercise professional must help the patient develop strategies to overcome these barriers to maintain physical activity as a regular part of daily life. Time stress, due to the sometimes overwhelming number and complexities of medical treatments and issues related to school demands, jobs, and family life, is a major barrier to achieving and maintaining physical activity habits. The exercise professional should help the patient to problem-solve ways to be time efficient without sacrificing time spent in physical activity. Some examples could include limiting screen time until after the physical activity is done, checking emails while riding a stationary bike, and incorporating bursts of activity into a workday, such as climbing stairs, parking further away from a store, or setting an alarm as a reminder to perform a brief bout of exercise.

For all of these recommendations, it is not essential that the physical activity be performed in a single bout. Breaking up the time into shorter intervals performed at different times in the day can improve adherence (eg, 10 minutes each in the morning, afternoon, and evening). Self-motivating tools such as step counters, accelerometers, and a wide variety of Internet tools or smart phone apps are available to provide reminders and help patients follow their physical activity plans. The exercise professional should investigate these options to recommend the best one(s) to help a particular patient at a particular time. It is also important to note that strategies to increase habitual physical activity may take 6 months or more to be effective in eliciting

behavior change,[35,36] so exercise professionals should strive for a long-term relationship with these patients.

Recommendations Regarding Aerobic Exercise for People With CF

Aerobic exercise is generally defined as physical activity that is performed continuously for at least 10 minutes that causes the heart rate and ventilation to increase.[11,18] There are many modes of aerobic exercise but most involve multiple large muscle groups working at the same time. Typical examples are walking, running, swimming, biking, and dancing. Recently, active video gaming systems have been shown to provide an adequate aerobic exercise stimulus for people with CF[37,38] and may be considered an alternative for some patients, especially those starting at very low levels of habitual physical activity. Aerobic exercise is the mode of exercise that has been most studied in people with CF and is now recommended for all patients by most CF centers.

The intensity of aerobic exercise is based on a percentage of the maximal capacity, ideally measured by an annual maximal exercise test.[6,39,107] Ideal intensity for aerobic training effect in CF is similar to healthy persons (70%–85% of maximal heart rate or 60%–80% of maximal oxygen consumption).[40] At this level, training-related improvements in cardiovascular, pulmonary, and metabolic systems will be seen but the intensity should be one that can be maintained for the 10 minutes or more duration. Intensity of exercise can be measured reliably in children with CF using the OMNI scale[41] or can be approximated by assessing the child's ability to carry on a conversation while still breathing noticeably harder.

Exercise intensity is important to consider. In a study of subjects with CF, it was found that moderate intensity (75% of maximal heart rate) was a better bronchodilator than albuterol; however, high-intensity exercise (90% of maximal heart rate) caused bronchoconstriction.[42] Similarly, studies have suggested that very intense exercise or exercise performed during pulmonary exacerbations may trigger a negative inflammatory response.[43]

Training effects have been seen equally in boys and girls with CF[44] and will be more pronounced in those patients who begin at a lower level of aerobic fitness.[45] Several studies have linked higher aerobic capacity with improved longevity[46] and participation in regular aerobic exercise slowed decline in forced vital capacity.[47] Recently, brief intervals of high-intensity training interspersed with active recovery periods have shown promise for increasing aerobic exercise capacity in adolescents and adults with severe CF lung dysfunction who were unable to perform standard continuous training.[48,49]

Clinical Question. How much aerobic exercise should be performed by a young child with CF?

Recommendation. Young children with CF should perform regular developmentally appropriate physical activity at least 60 minutes per day.

Evidence for Recommendation. Young children typically lack motor coordination to perform structured aerobic exercise. Also, they lack the cardiac and pulmonary development and also interest to sustain continuous physical activity. However, as the young child develops, walking, running, biking, swimming, and sport activities (soccer, basketball, etc.) are all effective activities for whole-body exercise. The young child will typically only perform these activities for short bursts (a few minutes at a time) and should perform a wide variety of activities to train motor patterns and encourage normal musculoskeletal development. Family involvement is a key at this stage, as is making exercise a play activity.

Clinical Question. How much aerobic exercise should be performed by a child with CF?

Recommendation. Aerobic exercise should be performed for 30–60 minutes 3 times per week or more to improve fitness. Up to 60 minutes per day of vigorous exercise can be helpful to meet physical activity guidelines.

Evidence for Recommendation. Based on a systematic review of training programs for children with CF, the intensity of aerobic exercise for this age group should be at least 70% of maximal heart rate to see improvements in lung function and maximal aerobic capacity.[50] Other outcomes seen in this age group include improved self-confidence with exercise and quality of life.[51] The duration of aerobic training programs for children with CF has ranged from 6 weeks to 3 years, and most have shown improvements in exercise capacity, hospitalization rates, and quality of life.[47,52–58] In patients who had very frequent pulmonary exacerbations, a targeted home program was shown to decrease hospitalization rate.[52] Both supervised and unsupervised exercise programs have been shown to improve exercise capacity; therefore, the exercise professional should consider the child's support system and attitudes toward exercise in determining the amount of supervision that is needed for an individual patient.

Children with CF should aim to begin more structured exercise programs as a way to meet the guidelines for moderate-to-vigorous physical activity. Children should be encouraged to seek out a variety of activities to suit their interests, such as taking classes in gymnastics; joining a recreational soccer, swimming, or basketball team; or participating in outdoor recreational clubs (eg, hiking or scouting). Participating as a family in a local fitness club can also be a way to instill regular exercise as part of the child's lifestyle. Children with CF will often cough more than their peers, and therefore it is important to educate coaches, physical education teachers, and parents that coughing is a desirable response. The exercise professional should be an advocate for including children with CF in all exercise outlets. Young children typically will self-limit their intensity by taking breaks or just slowing down, and exercise leaders should be educated that this behavior does not necessarily indicate

a lack of desire or motivation to participate. Although prepubertal children will not typically sweat excessively, the child with CF should be offered frequent water breaks and work with the CF center dietitian to develop strategies to prevent electrolyte or caloric depletion. Although many parents are concerned that participation in exercise will make their child lose weight, they should be informed that exercise is a known appetite stimulant and weight loss should not occur so long as nutritional intake is appropriate. In fact, a recent large study of self-reported exercise participation in children and adults with CF found that those who participated in exercise (even competitive sports) had higher body mass index (BMI) than those who did not exercise.[10] The CF center dietitian should work closely with patients, families, and the exercise professional to determine an individual diet plan to meet the increased demands of aerobic exercise.

Clinical Question. How much aerobic exercise should be performed by an adolescent with CF?

Recommendation. Aerobic exercise should be performed for 30–60 minutes 3 times per week or more to improve fitness. Up to 60 minutes per day of vigorous exercise can be helpful to meet physical activity guidelines.

Evidence for Recommendation. Adolescents are at high risk of becoming sedentary, and those with CF are no exception. Although the recommendation for aerobic exercise at this age group is based on sound evidence, it is also important to note that a short-term minimally supervised exercise program can be effective in helping adolescents to adopt physically active lifestyles that can persist up to 24 months.[59] It is important that the exercise professional set goals in conjunction with adolescents that allow them to experience success and benefits of exercise, even if they are not ready for the recommended duration or intensity. Patients who are sedentary will see improvements in fitness, energy level, and endurance more rapidly with lower exercise intensity than will those who are already regularly exercising. Participating in exercise can also improve postural problems that often begin to develop in adolescence.[60]

On the other end of the physical activity spectrum, adolescents with CF may be involved in team sports that require aerobic and other types of conditioning, and they should be encouraged to participate fully in this training. Most people will believe the need to expectorate secretions with high-intensity exercise, but those with CF will often produce more. It is important to educate coaches, physical education instructors, and teammates that coughing during exercise is desired and not to be feared. Most adolescents will recognize the need to take breaks and should be encouraged to work with their coaches to allow these as needed. People with CF can develop electrolyte abnormalities with excessive sweating, including exercise in the heat, and should work with the CF dietitian to develop strategies to replace electrolytes (such as consuming salty snacks or sports drinks) and to maintain adequate quantity and quality of caloric intake to meet the increased demands of exercise.

Adolescent patients should be counseled by exercise professionals about the benefits of aerobic exercise and the dangers of performance-enhancing substances that may be available to the patient. Participation in the same sports year round should be discouraged, as this could increase the risk of musculoskeletal injuries. Adolescents who express interest in extreme sports activities should be counseled to seek well-trained leaders who will teach techniques safely and not push them too quickly into high-level performance. The exercise professional needs to be aware of the popular exercise types and routines in general society and be prepared to analyze, along with the adolescent, the benefits and risks of their participation. Exercise professionals should also work with adolescents to develop plans to resume higher-level exercise after pulmonary exacerbations or injuries.

Note that The Exercise Working Group of the European Cystic Fibrosis Society recommends that all patients more than the age of 10 have regular maximal exercise tests to determine peak aerobic capacity.[107] This value has been shown to be a strong, and important, predictor of longevity in CF and has been recommended since at least 1971.[39] In addition, the results of this test will give the exercise professional critical information about any abnormal exercise responses that must be considered in the exercise prescription. Knowledge of a patient's current exercise capacity will also aid the exercise professional in setting goals to maintain or improve this level for the next year. It is important that the exercise professional understand how to interpret test outcomes to determine best modes of exercise for the individual patient. For example, a patient who is limited by ventilation may not be able to perform prolonged exercise using a large amount of muscle mass and should be guided toward interval training or resistance training as a viable alternative.

Clinical Question. How much aerobic exercise should be performed by an adult with CF?

Recommendation. Aerobic exercise should be performed for 30–60 minutes 3 times per week or more to improve aerobic fitness. Up to 60 minutes per day of aerobic exercise can be helpful to meet physical activity guidelines.

Evidence for Recommendation. Most evidence regarding the effects and benefits of exercise for people with CF has involved 30 minutes of training at moderate intensity 3–5 times per week for adolescents and young adults. These studies have shown improvements, or slower rate of decline, in lung function, BMD, rate of pulmonary exacerbation and hospitalizations, and quality of life. The most common modes of exercise have been treadmill walking and stationary bicycling, and training

has most commonly been performed for 4–6 weeks, although one study followed adults with CF for 1-year and saw improvements in lung function and exercise capacity compared with sedentary adults with CF.[61] Most of the initial benefits of aerobic training have, unfortunately, declined with the end of training, but longer-term studies have shown increased habitual physical activity levels, suggesting that participating in a structured training program led to adoption of exercise as a lifestyle.[59]

Complications of CF seen in adulthood are often improved with regular aerobic activity; however, considerations must be made for the demands of the particular exercise mode and intensity for patients with severe lung disease, severe malnutrition, poor bone density, joint pain, and metabolic issues such as CF-related diabetes (CFRD). Although most young adults with CF have physiology that allows them to adapt easily to the stresses of exercise, a slower approach may be needed with patients in middle age or later. Lower-impact activities may be more appropriate for those with joint and bone issues. Adults with CFRD must be aware of the effect of the exercise on blood glucose values and need to work closely with the CF dietitian and endocrinology team to develop a plan to manage blood glucose levels. Patients who are significantly underweight need to develop strategies to maximize caloric intake of appropriate quality to meet the additional demands of exercise. However, if sufficient calories are available, regular exercise may serve as an anabolic stimulus. A regular maximal exercise test will provide vital information to guide choice of exercise mode, intensity, and duration for each patient.

Many adults who are living well with CF attribute their success, at least in part, to a routine of regular aerobic exercise (with or without resistance training as discussed in Resistance Training Recommendations). Sharing their stories (readily available on Internet sites, such as YouTube and CFF.org) can help motivate adults with CF to adopt a physically active lifestyle. Exercise professionals need to work closely with these patients to adapt to their specific issues, both disease related (declining lung function, CFRD, and joint pains) and lifestyle related (job, home life, and financial situation) to help each patient be as physically active as possible throughout the lifespan.

Aerobic Exercise as Airway Clearance

There is keen interest from both patients and care team members about the use of aerobic exercise as an airway clearance modality. Growing evidence suggests that aerobic exercise can perform this role. Physical activities that demand higher ventilatory flows have been shown to increase liquid in the mucus of adults with CF,[33] and the impact of vigorous activities may loosen secretions from the airway walls, making expectoration easier.[7,62] In addition, there is evidence that moderate-intensity activity (80% of ventilatory threshold) can improve the nasal potential difference during exercise, which may, in turn increase water content of the mucus and decrease its viscosity.[63] However, other studies have shown that exercise airflows alone may not sufficiently move secretions without the addition of cough or huff techniques.[64] Therefore, patients should be advised to perform these techniques to fully clear secretions in addition to exercising.

Most children are able to learn methods of airway clearance that use different ventilatory volumes (eg, active cycle breathing and autogenic drainage). Aerobic exercise that causes the same ventilatory effects has been shown to be equally effective as more passive modes of airway clearance.[65] However, as airway clearance is needed daily, the exercise must be performed daily to be of benefit. In addition, the child must be willing to expectorate the mobilized secretions, such as pausing for huff or coughing, which can be distressing to children in a group environment. Exercise for 20 minutes at 60% of peak aerobic capacity has been found to decrease viscosity in secretions and improve ease of expectoration for adults with CF.[33]

Resistance Training Recommendations for People With CF

Strength is a vital part of function and necessary for many daily activities. Resistance training is the planned use of a load to increase the force output of a muscle contraction. It can be performed in a wide variety of ways, using body weight or equipment such as resistance bands or weights. It can also be performed in an aquatic environment, using the resistance of water.

Strength training is recommended for healthy children, adolescents, and adults as part of national guidelines.[11,66,67] In addition, strength training is specifically recommended for children and adolescents with CF by the Canadian Society for Exercise Physiology.[68]

Aside from improving ease of daily activities that require strength to perform, resistance training has the potential to improve disease-specific issues in CF. It is well known that resistance training can maintain or improve bone density and is routinely recommended for people at risk for osteoporosis. Resistance exercise can also increase lean body mass in both children and adults. It can be a way to improve mood and quality of life and can improve cardiovascular risk profile, which may be an emerging issue for middle-aged and older adults with CF.

It is important to note that just as aerobic training is prescribed at an intensity relative to maximal aerobic capacity, resistance training must be prescribed relative to maximal strength. The 1-repetition maximum test, in which a weight that can be lifted through a full movement using correct technique only one time, is the standard for strength testing. This technique has been used in several studies of children with CF and has been shown to be safe, if properly supervised.[53,56,57]

Clinical Question. What is the appropriate resistance training prescription for children and adolescents with CF?

Recommendation. Activities that develop muscular strength are recommended for all children. These include jumping, climbing, etc., and are part of normal developmental patterns beginning in toddlers. At an age where children can learn the required motor skills (typically age 7–8 years old[67]), formal resistance training can safely be performed, depending on the child/adolescent's interest, as part of a regular physical activity program. A full-body program should be prescribed, including multijoint movements. Large muscle groups should be trained before smaller groups. Resistance should begin with body weight (eg, push-ups, sit-ups, and pull-ups) and progress under skilled supervision to use of free weights or resistance bands. Plyometric activities (quick, explosive motions) should be incorporated as power training is important for sports and some daily activities. The following parameters are recommended for a formal training program:

- Frequency: 2–3 times per week
- Intensity: 50%–85% of 1-repetition maximum
- Volume: 1–3 sets of 6–15 repetitions.

Evidence for Recommendation. Resistance training is currently considered not only safe but also is recommended as part of a well-rounded physical activity plan for children, beginning at age 7–8, when motor and cognitive abilities begin to allow participation in formal, structured activities.[11,66] However, younger children should be participating in activities that lead to muscular strength using their own body weight. In 1984, Holzer et al[69] were the first to apply a program that included body weight strengthening exercises (ie, push-ups) to children with CF. Because of poor compliance, the program did not show significant changes, but this study is the first to determine that children with CF could perform these activities safely. In 1991, Horvat and Carlile[70] described systematic resistance training in 2 adolescents with CF and found improvements in strength, body composition, and self-concept. Selvadurai et al[56] and Orenstein et al[57] both performed randomized trials of programs including resistance training in children/youth. The resistance training group in the study of Orenstein et al[57] was found to improve not only strength of the trained muscle groups but also improved peak oxygen consumption. Similarly, Kriemler et al[59] found improvements in peak aerobic capacity and also in FEV_1 and hyperinflation after resistance training in patients aged 12 and above. In a 3-week program of combination aerobic and resistance training, Selvadurai et al[56] found improvements in FEV_1, leg strength, and lean body mass. Two systematic reviews[5,50] have supported the use of resistance training in children and adolescents with CF for improving strength outcomes. Recently, Urquhart et al[52] found improvements in hospital days, antibiotic use, shuttle walk test performance, and quality of life for a group of the most frequently hospitalized children who performed a combination aerobic and resistance training program. Santana Sosa et al[53] also found that a training program of both aerobic and resistance training improved not only strength of the trained muscles but also lean body mass and maximal oxygen consumption.

These findings certainly support that resistance training is safe for children and adolescents with CF, and most researchers also found improvements in strength and functional capacity. Therefore, the following considerations are made regarding resistance training in children and youth with CF.

Considerations for Children With CF. Any moderate or vigorous exertion can stimulate airway clearance and coughing; therefore, children should be encouraged to perform routine airway clearance before resistance training. Breathing patterns should be closely monitored, and children taught to avoid Valsalva and other breath-holding patterns. Nutritional intake should be monitored closely, with a goal toward maximizing protein and calcium intake and also overall caloric intake to support the activity.

Considerations for Adolescents With CF. As children reach puberty, hormone levels, particularly testosterone, will improve muscle hypertrophy gains with resistance training. Also, adolescents typically have the necessary motor control and maturity to learn more complex resistance training movements. Adolescents involved in sports will have specific needs for strength dependent on the sport. Thus, the supervising exercise specialist should consider the individual's needs in designing a specific program. However, the parameters listed above for children should be continued in adolescence as part of a healthy physical activity program.

In addition, care should be taken to determine any joints at risk for loss of flexibility and exercises designed to target these areas. For example, exercises that strengthen scapular retractor muscles will help minimize the effect of air trapping on development of a rounded shoulder posture. Exercises that provide weight bearing through long bones and spine are encouraged during this time of maximal bone accrual to help prevent osteoporosis in adulthood.

Clinical Question. What is the appropriate resistance training program for adults with CF?

Recommendation.

- Frequency: 2–3 times per week
- Intensity: at least 70% of 1-repetition maximum
- Volume: 1–3 sets of 8–15 repetitions
- Type: a program that includes upper body, lower body, and trunk muscles is recommended. Multijoint motions using large muscle groups should be performed before small or single-joint motions. Resistance can include body weight, resistance bands, or weight equipment.

Evidence for Recommendation. Resistance training is recommended for healthy adults by the American College of Sports Medicine (ACSM)[11] and World Health

Organization.[18] It is also recommended for people with many chronic diseases, such as arthritis, osteoporosis, diabetes, heart disease, and stroke. Optimal outcomes include maintaining or improving ability to perform daily activities, improving cardiovascular disease risk, and improving body composition. Because people with CF live longer into adulthood, issues related to metabolism, bone health, and perhaps cardiovascular health will become more prevalent. It is important, then, to make sure adults with CF perform a well-rounded physical activity program.

Resistance training was first examined in young adults by Strauss et al[71] in 1987 who found that a variable weight training program using a pyramidized program improved strength, muscle size (hypertrophy), body weight, and pulmonary function. However, since then, little study of resistance training in adults has been undertaken. Many exercise programs including older adolescents (ages 16–21) have involved a combination of aerobic and resistance training; thus, determining effects of resistance training alone has been difficult. Recently, Kriemler et al[59] performed a study in which young adults with CF were randomized to aerobic or resistance training groups. The groups were supervised for 6 months and then continued independently to 12 months. Although the intensity of the resistance training was not stated, a full-body program was designed. At the end of the intervention, the resistance training group showed improvements in FEV_1 and forced vital capacity (FVC) that were similar to the aerobic training group, and also decreased hyperinflation. These findings suggest that resistance training may be as beneficial for lung function as an aerobic training program. Because not all people enjoy aerobic activities, it is good to know that resistance training has the potential to benefit lung function in people with CF.

Considerations for Adults With CF. For people with low bone density, care should be taken to avoid ballistic movements and motions that involve vigorous trunk flexion or rotation. For people who develop CFRD, blood glucose monitoring should be performed before, during, and after exercise. Nutritional advice from the CF Center dietitian should be sought to ensure not only adequate caloric intake to balance the expenditure of exercise but also maximizing protein and calcium intake. Proper breathing, especially avoiding breath-holding, should be encouraged. Exercises that provide load through axial spine and legs should be encouraged to minimize bone loss. Stretching to reduce or eliminate typical muscular tightness (eg, rounded shoulders and forward head posture) should be incorporated along with resistance exercises to strengthen antagonist muscles (eg, scapular retractors).

Special Situations for Resistance Training in CF

Peripheral muscle weakness has been noted in both adolescents and adults with CF.[72–78] Although the cause of the weakness is unclear, some studies have indicated that people with CF may have metabolic abnormalities in peripheral muscle, which impair ability to produce adenosine triphosphate (ATP) to generate muscle contraction and/or impair replenishment of ATP and creatine phosphate after exercise.[79–83] These abnormalities may translate to a need for longer rest periods between repetitions of resistance training exercise or perhaps longer rest periods between training days. As resistance training has not been widely studied, particularly in adults with CF, the optimal rest period has not been determined.

For people with severe lung disease, resistance training may be an optimal mode of exercise, as there is lower ventilatory demand than with continuous aerobic exercise. The use of neuromuscular electrical stimulation and more traditional resistance training has been recommended to help moderate declines in muscle strength associated with pulmonary exacerbations in people with chronic lung diseases.[84]

The use of corticosteroids[85] and antirejection medications taken after lung transplantation[86] has been shown to impair muscle size and strength, which can lead to significant limitations to daily activity and exercise tolerance. Thus, targeted muscle training (such as resistance training) should be part of the rehabilitation plan after acute exacerbation and particularly after lung transplantation to counter the anticipated decline in function.[87] It is important that the rehabilitation professionals understand the influence of pretransplant limitations, concomitant diseases (eg, CFRD, hypertension, and osteoporotic fractures), and nutritional status on the patient's ability to perform exercise training. In cases of transplantation or severe CF disease complications, an exercise program of resistance training must be individualized with the goal to reach the recommended levels stated previously for children or adults with CF who do not have these complications.

Patients should be cautioned not to use "performance enhancing" supplements commonly used by people who wish to gain weight, increase muscle mass, or enhance performance, without first discussing these products with the CF care center physician and/or dietitian.

Precautions/Contraindications for Exercise and Habitual Physical Activity for People With CF

Despite the potential disease-related effects on exercise ability, exercise and physical activity have been shown to be quite safe for people with CF.[88] Adverse events such as cardiac arrhythmias, pneumothorax, and hypoglycemia associated with exercise occurred in less than 1% of nearly 1000 people with CF surveyed in 37 centers.[88] However, exercise specialists may be faced with questions regarding exercise and physical activity for patients with known CF complications. In this section, guidance is provided for many of these situations.

Clinical Question. Should patients with CF exercise using supplemental oxygen?

Patients who desaturate during physical activity or exercise should receive appropriate levels of oxygen supplementation to maintain oxygen saturation levels above 90% during exercise.[89,90] This is important, not only to improve exercise performance and recovery[91–94] but also to help prevent development of pulmonary hypertension and/or cor pulmonale.

Clinical Question. What physical activity considerations are there for patients with CF after lung transplantation?

A comprehensive exercise program after lung transplantation should include aerobic, strength, and flexibility components and also neuromuscular and functional activities, and begin and end with a warm up and cool down.[95] Because of the effects of glucocorticoids for immunosuppression[95] and other peripheral limitations to exercise,[96] strength training for peripheral muscles should be a particular focus. It would be prudent to determine the patient's history of osteoporosis and fractures and focus on strengthening and weight-bearing exercises to combat osteoporosis in the vertebrae and hips,[97,98] as this is a common comorbidity in patients with lung transplant, especially in the presence of CFRD.[89] During exercise sessions, rating of perceived exertion and dyspnea rating scales should be used to monitor intensity because many patients do not reach targeted heart rates.[95] A drop in oxygen saturation levels and other signs of exercise intolerance, such as excessive or increasing shortness of breath, should be reported to the transplant team, as these can indicate organ rejection or infection.[95] Special care should be taken to clean exercise equipment before use by patients taking immunosuppressive medications because of increased risk of developing infections.[95] For more detailed information about exercise guidelines before and after lung transplant, see the study by Hirche et al.[87]

Clinical Question. Should patients with CF-related diabetes perform exercise/physical activity?

Continuing with physical activity is recommended for patients with CFRD even though it is likely to make physical activity more difficult to perform. Additional precautions should be used to monitor blood glucose, especially after exercise, even though the occurrence of hypoglycemia is low.[88] Pulmonary function and BMI should be monitored in this population because of the association of these measures with CFRD.[89] Because CFRD affects bone health, recommendations for people with low bone density should also be followed.[99]

Clinical Question. What are the physical activity recommendations for patients with CF who have osteopenia or osteoporosis?

Everyone with CF should be encouraged to participate in regular physical activity as this has been shown to be associated with greater BMD.[100] Determine the history of fractures in patients with CF.[101] Because low BMI is related to bone loss, BMI should be followed regularly to screen for referral to a dietician; the target should be a BMI greater than 50 percentile.[99] More research is needed to define optimal physical activity programs to enhance BMD, but experts encourage weight-bearing activity.[89,99] Lower extremity strength training programs may be especially beneficial for low BMD in the proximal femur.[97]

Clinical Question. How soon after hemoptysis should physical activity/exercise be started/resumed?

There are no current guidelines regarding specific timing and types of exercise after hemoptysis, a fairly common complication of CF. In an informal survey of over 50 CF centers worldwide, hemoptysis was defined as mild (<5 mL blood), moderate (5–240 mL), or severe (>240 mL). Most physicians surveyed would recommend temporary cessation of exercise after a moderate or severe bleed. Gradual resumption of the intensity of routine exercise is also recommended after 24–48 hours of no new bleeding.[90] Australian guidelines[102] also recommend limiting heart rate rise during this acute posthemoptysis period.

Clinical Question. What are the recommendations for physical activity in the patient with CF and pneumothorax?

The possibility of pneumothorax should be considered for adults and patients with poor lung function,[89,103] and also the high likelihood for recurrence in patients with a history.[104] It is prudent to advise airway clearance be performed before exercise to reduce unnecessary coughing, as exercise-related coughing has been reported to cause pneumothorax.[88] Even without concrete evidence demonstrating a direct link between pneumothorax and exercise, an expert consensus panel recommends that patients with pneumothorax of any size should "not lift weights greater than 5 lbs for 2 weeks after the pneumothorax has resolved."[103] An informal survey of CF centers worldwide has indicated that a limitation on vigorous exercise or high-load weight lifting would be avoided in the acute recovery period, because of concerns that Valsalva maneuver could slow healing or expand a pneumothorax. In addition, most CF centers would advise against scuba diving in all people with CF and would be very cautious about high-altitude activities for patients with risk of pneumothorax development (eg, those with severe lung disease). Concerns about pneumothorax with vigorous coughing, which may be incurred during exercise, should also encourage patients to use airway clearance techniques that minimize this risk (eg, huff coughing). A gradual resumption of upper body exercise over 2–4 weeks after removal of a chest tube for pneumothorax is recommended.[90]

Clinical Question. What type of physical activity is appropriate for people with CF who have poor nutrition and/or low body mass index?

Physical activity advice should be related to energy requirements of the chosen activity. A dietitian should be closely involved to determine individual energy requirements and consult on dietary changes to accompany an exercise program.[89] The expert consensus is that exercise is rarely, if ever, a cause of weight loss as long as dietary

intake is sensibly managed. In fact, people with CF who participated in exercise were found to have higher BMI than those who did not.[10]

Clinical Question. What type of exercise is appropriate for patients with CF during/after pregnancy?

For pregnant women, efforts at physical activity are complicated by changes in pregnancy; these changes could present an even larger problem for a woman with CF. Although it is not necessary to avoid exercise during pregnancy and in the postpartum period, exercise intensity should be decreased as the pregnancy advances. Pulmonary function and nutrition status should be closely followed in women with CF during and after pregnancy. This is also an important time to reinforce teaching pelvic floor muscle exercises to help with postdelivery stress urinary incontinence.[102]

Clinical Question. Is physical activity still considered prudent during a pulmonary exacerbation or inpatient hospital stay?

It is important that some kind of exercise be performed in the acute care setting, even in the intensive care unit, to continue to accrue multiple benefits of physical activity. Exercise intensity should be decreased during an exacerbation, as patients will need less effort to reach an age-related target heart rate.[105] Patients hospitalized with an exacerbation should receive greater monitoring and a specific exercise program based on individual vital signs and overall response to physical activity.

Clinical Question. What are the precautions for exercise in patients with indwelling vascular or nutritional catheters (eg, peripherally inserted central [PICC] lines, ports, and J-tubes)?

No specific guidelines exist for exercise in the presence of indwelling devices; however, contact sports or activities where trauma to the site of the device might occur should be avoided. Vigorous use of the upper arm containing a peripherally inserted central catheter should also be limited to avoid tension on the catheter.

Clinical Question. Are there activities that should not be performed by people with CF?

Most experts agree that scuba diving and mountaineering at high altitude pose risks to people with CF and should be avoided. The risk is related to changes in air pressure (diving) or ambient pressure of oxygen (altitude). These activities could lead to pneumothorax or high-altitude pulmonary edema. In addition, patients who have impaired clotting function (eg, CF liver disease and treatment with anticoagulant medications) should avoid contact sports, or those with high potential for trauma (eg, horse jumping, bungee jumping, and military style high-intensity training programs).

CONCLUSIONS

Physical activity and exercise should be part of the routine management for people with CF at any age. This document provides guidance to the CF care team for counseling patients regarding types and amounts of physical activity to meet recommended goals. Guidelines for adapting physical activity and exercise for common CF-related conditions have also been provided. The ultimate goal of this document is to increase the number of patients who adopt a physically activity lifestyle for health and good quality of life.

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REFERENCES

1. CF Foundation. About CF. www.cff.org. Accessed May 15, 2015.
2. Cystic Fibrosis Canada. Median age of survival surpasses 50 years of age for Canadians with cystic fibrosis. News and media web site. 2014. http://www.cysticfibrosis.ca/median-age-of-survival-surpasses-50-years-of-age-for-canadians-with-cystic-fibrosis/. Accessed June 15, 2015.
3. Boas SR. Exercise recommendations for individuals with cystic fibrosis. *Sports Med.* 1997;24:17–37.
4. Bradley JM, Moran F. Physical training for cystic fibrosis. *Cochrane Database Syst Rev.* 2008(1):CD002768.
5. Shoemaker MJ, Hurt H, Arndt L. The evidence regarding exercise training in the management of cystic fibrosis: a systematic review. *Cardiopulm Phys Ther J.* 2008;19:75–83.
6. Rand S, Prasad SA. Exercise as part of a cystic fibrosis therapeutic routine. *Expert Rev Respir Med.* 2012;6:341–352.
7. Wheatley CM, Wilkins BW, Snyder EM. Exercise is medicine in cystic fibrosis. *Exerc Sport Sci Rev.* 2011;39:155–160.
8. Cerny F. Exericse and cystic fibrosis (CF) 2.0. *Pediatr Exerc Sci.* 2013; 25:616–623.
9. LeBlanc CMA, Lands LC. Can I play? *Pediatr Ann.* 2014;43:e316–e324.
10. Collaco JM, Blackman SM, Raraigh KS, et al. Self-reported exercise and longitudinal outcomes in cystic fibrosis: a retrospective cohort study. *BMC Pulm Med.* 2014;14:159.
11. American College of Sports Medicine. *ACSM's Guidelines for Exercise Testing and Prescription.* 9th ed . Baltimore, MD: Wolters Kluwer; 2014.
12. Cropp GJ, Pullano TP, Cerny FJ, et al. Exercise tolerance and cardiorespiratory adjustments at peak work capacity in cystic fibrosis. *Am Rev Respir Dis.* 1982;126:211–216.
13. Urquhart DS, Odendaal D, Derrick G, et al. Exercise prescription in CF—a therapeutic modality for all? *Pediatr Pulmonol.* 2006;41(S29):354.
14. Lamhonwah A, Bear C, Huan L, et al. Cystic fibrosis transmembrane conductance regulator in human muscle: dysfunction causes abnormal metabolic recovery in exercise. *Ann Neurol.* 2010;67:802–808.

15. Dekerlegand RL, Hadjiliadis D, Swisher AK, et al. Inspiratory muscle strength relative to disease severity in adults with stable cystic fibrosis. *J Cyst Fibros.* In press.
16. Poore S, Berry B, Eidson D, et al. Evidence of vascular endothelial dysfunction in young people with cystic fibrosis. *Chest.* 2013;143:939–945.
17. Schrage W, Wilkins B, Dean J, et al. Exercise hyperemia and vasoconstrictor responses in humans with cystic fibrosis. *J Appl Physiol (1985).* 2005;99:1866–1871.
18. World Health Organization. Global Recommendations on Physical Activity for Health. 2011. http://www.who.int/dietphysicalactivity/pa/en/index.html. Accessed July 15, 2014.
19. Nixon P, Orenstein D, Kelsey S. Habitual physical activity in children and adolescents with cystic fibrosis. *Med Sci Sports Exerc.* 2001;33(1):30–35.
20. Selvadurai H, Blimkie C, Cooper P, et al. Gender differences in habitual activity in children with cystic fibrosis. *Arch Dis Child.* 2004;89:928–933.
21. Boucher G, Lands L, Hay J, et al. Activity levels and the relationship to lung function and nutritional status in children with cystic fibrosis. *Am J Phys Med Rehabil.* 1997;76(4):311–315.
22. Loutzenhiser J, Clark R. Physical activity and exercise in children with cystic fibrosis. *J Pediatr Nurs.* 1993;8(2):112–119.
23. Stanghelle J, Hjeltnes N, Bangstad H, et al. Effect of daily short bouts of trampoline exercise during 8 weeks on the pulmonary function and the maixmal oxygen uptake of children with cystic fibrosis. *Int J Sports Med.* 1988;9(suppl 1):32–36.
24. Managing Nutrition & Digestive Problems: Exercise and CF. In: *Cystic Fibrosis Family Education Program*. Bethesda, MD: Cystic Fibrosis Foundation; 2013:1–6.
25. Britto M, Garret J, Konrad T, et al. Comparison of physical activity in adolescents with cystic fibrosis versus age-matched controls. *Pediatr Pulmonol.* 2000;30(2):86–91.
26. Schneiderman-Walker J, Wilkes D, Strug L, et al. Sex differences in habitual physical activity and lung function decline in children with cystic fibrosis. *J Pediatr.* 2005;147:321–326.
27. Schneiderman JE, Wilkes DL, Atenafu EG, et al. Longitudinal relationship between physical activity and lung health in patients with cystic fibrosis. *Eur Respir J.* 2014;43:817–823.
28. Hebestreit H, Keiser S, Junge S, et al. Long-term effects of a partially supervised conditioning programme in cystic fibrosis. *Eur Respir J.* 2010;35:578–583.
29. Hebestreit H, Kieser S, Rudiger S, et al. Physical activity is independently related to aerobic capacity in cystic fibrosis. *Eur Respir J.* 2006;28:734–739.
30. Troosters T, Langer D, Vrijsen B, et al. Skeletal muscle weakness, exercise tolerance and physical activity in adults with cystic fibrosis. *Eur Respir J.* 2009;33:99–106.
31. Ward N, White D, Rose H, et al. Physical activity levels of patients with cystic fibrosis hospitalised with an acute respiratory exacerbation. *Respir Med.* 2013;107:1014–1020.
32. Rasekaba TM, Button BM, Wilson JW, et al. Reduced physical activity associated with work and transport in adults with cystic fibrosis. *J Cyst Fibros.* 2013;12:229–233.
33. Dwyer TJ, Alison JA, McKeorgh ZJ, et al. Effects of exercise on respiratory flow and sputum properties in patients with cystic fibrosis. *Chest.* 2011;139:870–877.
34. Moola FJ, Faulkner GE, Schneiderman JE. "No time to play": perceptions toward physical activity in youth with cystic fibrosis. *Adapt Phys Activ Q.* 2012;29:44–62.
35. Cox NS, Alison JA, Holland AE. Interventions for promoting physical activity in people with cystic fibrosis. *Cochrane Database Syst Rev.* 2013;12:CD009448.
36. Hommerding PX, Baptista RR, Makarewicz GT, et al. Effects of an educational intervention of physical activity for children and adolescents with cystic fibrosis: a randomized controlled trial. *Respir Care.* 2015;60:81–87.
37. Kuys SS, Hall K, Peasey M, et al. Gaming console exercise and cycle or treadmill exercise provide similar cardiovascular demand in adults with cystic fibrosis: a randomised cross-over trial. *J Physiother.* 2011;57:35–40.
38. Salonini E, Gambazza S, Meneghalli I, et al. Active video game playing in children and adolescents with cystic fibrosis: exercise or just fun? *Respir Care.* 2015;60(8):1172–1179.
39. Godfrey S, Mearns M. Pulmonary function and response to exercise in cystic fibrosis. *Arch Dis Child.* 1971;46:144–151.
40. Williams CA, Benden C, Stevens D, et al. Exercise training in children and adolescents with cystic fibrosis: theory into practice. *Int J Pediatr.* 2010;2010. doi: 10.1155/2010/670640.
41. Higgins LW, Robertson RJ, Kelsey SF, et al. Exercise intensity self-regulation using the OMNI scale in children with cystic fibrosis. *Pediatr Pulmonol.* 2013;48:497–505.
42. Wheatley CM, Baker SE, Morgan MA, et al. Effects of exercise intensity compared to albuterol in individuals with cystic fibrosis. *Respir Med.* 2015;109:463–474.
43. van de Weert-van Leeuwen PB, Slieker MG, Hulzebos HJ, et al. Chronic infection and inflammation affect exercise capacity in cystic fibrosis. *Eur Respir J.* 2012;39:893–898.
44. Gruber W, Orenstein DM, Braumann KM, et al. Effects of an exercise program in children with cystic fibrosis: are there differences between females and males? *J Pediatr.* 2011;158:71–76.
45. Gruber W, Orenstein DM, Braumann KM. Do responses to exercise training in cystic fibrosis depend on initial fitness level? *Eur Respir J.* 2011;38:1336–1342.
46. Nixon P, Orenstein D, Kelsey S, et al. The prognostic value of exercise testing in patients with cystic fibrosis. *N Engl J Med.* 1992;327(25):1785–1788.
47. Schneiderman-Walker J, Pollock S, Corey M, et al. A randomized controlled trial of a 3-year home exercise program in cystic fibrosis. *J Pediatr.* 2000;136:304–310.
48. Gruber W, Orenstein DM, Braumann KM, et al. Interval exercise training in cystic fibrosis—effects on exercise capacity in severely affected adults. *J Cyst Fibros.* 2014;13:86–91.
49. Hulzebos H, Snieder H, van der Net J, et al. High-intensity interval training in an adolescent with cystic fibrosis: A physiological perspective. *Physiother Theory Pract.* 2011;27(3):231–237.
50. van Doorn N. Exercise programs for children with cystic fibrosis: a systematic review of randomized controlled trials. *Disabil Rehabil.* 2010;32:41–49.
51. Gulmans V, de Meer K, Brackel H, et al. Outpatient exercise training in children with cystic fibrosis: physiological effects, perceived competence, and acceptability. *Pediatr Pulmonol.* 1999;28:39–46.
52. Urquhart D, Sell Z, Dhouieb E, et al. Effects of a supervised, outpatient exercise and physiotherapy programme in children with cystic fibrosis. *Pediatr Pulmonol.* 2012;47:1235–1241.
53. Santana Sosa E, Groeneveld IF, Gonzalez-Saiz L, et al. Intrahospital weight and aerobic training in children with cystic fibrosis: a randomized controlled trial. *Med Sci Sports Exerc.* 2012;44(1):2–11.
54. Elbasan B, Tunali N, Duzgun I, et al. Effects of chest physiotherapy and aerobic exercise training on physical fitness in young children with cystic fibrosis. *Ital J Pediatr.* 2012;38(2). http://www.ijponline.net/content/38/1/2.
55. Santana-Sosa E, Gonzalez-Saiz L, Groeneveld IF, et al. Benefits of combining inspiratory muscle with "whole muscle" training in children with cystic fibrosis: a randomised controlled trial. *Br J Sports Med.* 2014;48(20):1513–1517.
56. Selvadurai H, Blimkie C, Meyers N, et al. Randomized controlled study of in-hospital exercise training programs in children with cystic fibrosis. *Pediatr Pulmonol.* 2002;33:194–200.
57. Orenstein D, Hovell M, Mulvihill M, et al. Strength vs aerobic training in children with cystic fibrosis. *Chest.* 2004;126:1204–1214.
58. Hebestreit H, Schmid K, Kieser S, et al. Quality of life is associated with physical activity and fitness in cystic fibrosis. *BMC Pulm Med.* 2014;14:26.
59. Kriemler S, Keiser S, Junge S, et al. Effect of supervised training on FEV_1 in cystic fibrosis: a randomised controlled trial. *J Cyst Fibrosis.* 2013;12(6):714–720.
60. Schindel CS, Hommerding PX, Melo DAS, et al. Physical exercise recommendations improve postural changes found in children and adolescents with cystic fibrosis: a randomized controlled trial. *J Pediatr.* 2015;166:710–716.

61. Moorcroft A, Dodd M, Morris J, et al. Individualised unsupervised exercise training in adults with cystic fibrosis: a 1 year randomised controlled trial. *Thorax*. 2004;59:1074–1080.
62. Dwyer TJ, Elkins MR, Bye PTP. The role of exercise in maintaining health in cystic fibrosis. *Curr Opin Pulm Med.* 2011;17:455–460.
63. Hebestreit A, Kersting U, Basler B, et al. Exercise inhibits epithelial sodium channels in patients with cystic fibrosis. *Am J Respir Crit Care Med.* 2001;164:443–446.
64. McIlwaine MP, Son NML, Richmond ML. Physiotherapy and cystic fibrosis: what is the evidence base? *Curr Opin Pulm Med.* 2014;20: 613–617.
65. Cerny F. Relative effects of bronchial drainage and exercise for in-hospital care of patients with cystic fibrosis. *Phys Ther.* 1989;69: 633–639.
66. Lloyd RS, Faigenbaum AD, Stone MH, et al. Position statement on youth resistance training: the 2014 International Concensus. *Br J Sports Med.* 2014;48(7):498–505.
67. Faigenbaum AD, Kraemer WJ, Blimkie CJR, et al. Youth resistance training: updated position statement paper from the National Strength and Conditioning Association. *J Strength Cond Res.* 2009;23:s60–s79.
68. Behm DG, Faigenbaum AD, Falk B, et al. Canadian Society for Exercise Physiology position paper: resistance training in children and adolescents. *Appl Physiol Nutr Metab.* 2008;33:547–561.
69. Holzer F, Schnall R, Landau L. The effect of a home exercise programme in children with cystic fibrosis and asthma. *Aust Paedriatr J.* 1984;20:297–302.
70. Horvat M, Carlile JR. Effects of progressive resistance exercise on physical functioning and self-concept in cystic fibrosis. *Clin Kinesiol.* 1991;45:18–23.
71. Strauss G, Osher A, Chun-I W, et al. Variable weight training in cystic fibrosis. *Chest*. 1987;92:273–276.
72. Lands L, Heigenhauser G, Jones N. Respiratory and peripheral muscle function in cystic fibrosis. *Am Rev Respir Dis.* 1993;147:865–869.
73. de Meer K, Gulmans V, van der Laag J. Peripheral muscle weakness and exercise capacity in children with cystic fibrosis. *Am J Respir Crit Care Med.* 1999;159:748–754.
74. de Jong W, van Aalderen W, Kraan J, et al. Skeletal muscle strength in patients with cystic fibrosis. *Physiother Theory Pract.* 2001;17:23–28.
75. Hussey J, Gormley J, Leen G, et al. Peripheral muscle strength in young males with cystic fibrosis. *J Cyst Fibros.* 2002;1:116–121.
76. Pinet C, Cassart M, Scillia P, et al. Function and bulk of respiratory and limb muscles in patients with cystic fibrosis. *Am J Respir Crit Care Med.* 2003;168:989–994.
77. Swisher A. Not just a lung disease: peripheral skeletal muscle abnormalities in cystic fibrosis and the role of exercise to address them. *Cardiopulm Phys Ther J.* 2006;17(1):10–15.
78. Swisher AK, Baer L, Moffett K. The influence of lean body mass and leg muscle strength on 6-minute walk test performance in children with cystic fibrosis. *Cardiopulm Phys Ther J.* 2005;16:5–9.
79. deMeer K, Jeneson J, Gulmans V, et al. Efficiency of oxidative work performance of skeletal muscle in patients with cystic fibrosis. *Thorax*. 1995;50:980–983.
80. Moser C, Tirakitsoontorn P, Nussbaum E, et al. Muscle size and cardiorespiratory response to exercise in cystic fibrosis. *Am J Respir Crit Care Med.* 2000;162:1823–1827.
81. Selvadurai H, Allen J, Sachinwalla T, et al. Muscle function and resting energy expenditure in female athletes with cystic fibrosis. *Am J Respir Crit Care Med.* 2003;168:1476–1480.
82. Sahlberg M, Svantesson U, Magnusson Thomas E, et al. Muscular strength and function in patients with cystic fibrosis. *Chest*. 2005;127: 1587–1592.
83. Wells G, Wilkes D, Schneiderman J, et al. Skeletal muscle metabolism in cystic fibrosis and primary ciliary dyskinesia. *Pediatr Res*. 2011;69: 40–45.
84. Spruit MA, Singh SJ, Garvey C, et al. An official American Thoracic Society/European Respiratory Society statement: key concepts and advances in pulmonary rehabilitation. *Am J Respir Crit Care Med.* 2013;188:e13–e64.
85. Barry S, Gallagher C. Corticosteriods and skeletal muscle function in cystic fibrosis. *J Appl Physiol (1985).* 2003;95:1379–1384.
86. Pinet C, Scillia P, Cassart M, et al. Preferential reduction of quadriceps over respiratory muscle strength and bulk after lung transplantation for cystic fibrosis. *Thorax*. 2004;59:783–789.
87. Hirche TO, Knoop C, Hebestreit H, et al. Practical guidelines: lung transplantation in patients with cystic fibrosis. *Pulm Med.* 2014;2014:621342.
88. Ruf K, Winkler B, Hebestreit A, et al. Risks associated with exercise testing and sports participation in cystic fibrosis. *J Cyst Fibros*. 2010;9: 339–345.
89. Yankaskos JR, Marshall BC, Sufian B. Cystic fibrosis adult care-consensus conference report. *Chest*. 2004;125:S1–S39.
90. Burtin C, Hebestreit H. Rehabilitation in patients with chronic respiratory disease other than chronic obstructive pulmonary disease: exercise and physical activity interventions in cystic fibrosis and non-cystic fibrosis bronchiectasis. *Respiration*. 2015;89:181–189.
91. Marcus CL, Bader D, Stabile MW, et al. Supplementary oxygen and exercise performance in patients with cystic fibrosis with severe pulmonary disease. *Chest*. 1992;101:52–57.
92. McKone E, Barry S, FitzGerald M, et al. Role of arterial hypoxemia and pulmonary mechanics in exercise limitation in adults with cystic fibrosis. *J Appl Physiol (1985).* 2005;99:1012–1018.
93. Nixon PA, Orenstein DM, Curtis SE, et al. Oxygen supplementation during exercise in cystic fibrosis. *Am Rev Respir Dis.* 1990;142:807–811.
94. Shah AR, Keens TG, Gozal D. Effect of supplemental oxygen on supramaximal exercise performance and recovery in cystic fibrosis. *J Appl Physiol (1985).* 1997;83:1641–1647.
95. Balfe DL, Ross DJ. Lung and heart-lung transplantation. In: Durstine JL, More GE, Painter PL, et al, eds. *ACSM's Exercise Management for Persons with Chronic Diseases and Disabilities*. 3rd ed . Champaign, IL: Human Kinetics; 2009.
96. Reid MS, Levy RD. Exercise limitation in recipients of lung transplants. *Phys Ther.* 2004;84:1178–1187.
97. Dodd JD, Barry SC, Barry RBM. Bone mineral density in cystic fibrosis: benefit of exercise capacity. *J Clin Densitom.* 2008;11:537–542.
98. Michell MJ. Resistance training prevents vertebral osteoporosis in lung transplant recipients. *Transplantation*. 2003;76:557–762.
99. Gore AP, Kwon SH, Stenbit AP. A roadmap to the brittle bones of cystic fibrosis. *J Osteoporos.* 2011. doi:10.4061/2011/926045.
100. Garcia ST, Giraldez Sanchez MA, Cejudo P, et al. Bone health, daily physical activity, and exercise tolerance in patients with cystic fibrosis. *Chest*. 2011;140:475–481.
101. Aris R, Merkel P, Bachrach L, et al. Guide to bone health and disease in cystic fibrosis. *J Clin Endocrinol Metab.* 2005;90:1888–1896.
102. Holland AE. Physiotherapy for cystic fibrosis in Australia: A consensus statement. 2008. http://www.thoracic.org.au/documents.html. Accessed June19, 2015.
103. Flume PA, Mogayzel PJ, Robinson KA. Cystic fibrosis pulmonary guidelines. Pulmonary complications: hemoptysis and pneumothorax. *Am J Respir Crit Care Med.* 2010;182:298–305.
104. Stenbit A, Flume PA. Pulmonary complications in adult patients with cystic fibrosis. *Am J Med Sci.* 2008;335:55–59.
105. Nixon PA. Cystic Fibrosis. In: Durstine JL, More GE, Painter PL, et al, eds. *ACSM's Exercise Management for Persons with Chronic Diseases and Disabilities*. 3rd ed . Champaign, IL: Human Kinetics; 2009.
106. Bradley J, O'Neill B, Kent L, et al. Physical activity assessment in cystic fibrosis: a position statement [published online ahead of print July 25, 2015]. *J Cyst Fibros*. doi:10.1016/j.jcf.2015.05.011.
107. Hebestreit H, Arets H, Aurora P, et al. Statement on exercise testing in cystic fibrosis. *Respiration*. In Press.