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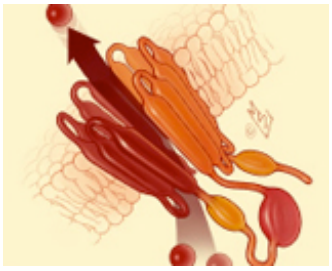

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EMERGING CF THERAPIES 2009

eCysticFibrosis Review Volume 3 Issue 7

The Role of Exercise and Physical Activity in Optimizing Outcomes Among Patients With Cystic Fibrosis

In this Issue...

Despite advances in clinical care, life expectancy in patients with cystic fibrosis (CF) remains shortened. It has been well documented that exercise capacity in this population is limited by impaired lung function, peripheral skeletal muscle function, and nutritional status, as well as the inability of the cardiorespiratory system to meet the metabolic demands associated with exercise. Interestingly, the physiologic consequences observed in patients with CF are similar to the effects of deconditioning, including poor cardiovascular function, reduced muscle mass, and impaired strength and power. Further, children with CF may be more physically inactive because of the burden of their chronic disease and thus may be at risk for compounding the combined effects of chronic disease and physical inactivity. Exercise and physical activity are key factors in the management of patients with CF, as such markers of physical fitness as aerobic capacity are related to pulmonary function and may be associated with mortality.

In this issue, we review the pathophysiology of exercise intolerance in patients with CF, and discuss new research on how clinicians can incorporate physical activity into the treatment plan for their patients with CF, with the expectation that exercise can improve outcomes in this population.

LEARNING OBJECTIVES

After participating in this activity, the participant will demonstrate the ability to:

- Describe the pathophysiology of exercise intolerance in patients with cystic fibrosis (CF)
- Discuss the research on the relationship between exercise function/capacity and longitudinal clinical outcomes in patients with CF
- Summarize how to implement exercise and physical activity recommendations to optimize clinical outcomes in patients with CF

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IN THIS ISSUE

GUEST AUTHOR OF THE MONTH

In This Issue & Reviews:

Greg D. Wells, PhD
Associate Scientist in Physiology and Experimental Medicine
The Hospital for Sick Children
Toronto, Ontario, Canada
Assistant Professor
Faculty of Kinesiology and Physical Education
Director
Human Physiology Research Unit
University of Toronto
Toronto, Ontario, Canada

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COMMENTARY

Long-term prognosis and survival remain significant issues among patients with cystic fibrosis (CF), despite advances in treatment and care.[1] Exercise capacity in this group is limited by impaired pulmonary function, peripheral skeletal muscle function, and nutritional status, as well as by the capacity of the cardiorespiratory system to endure the metabolic demands of exercise.[2] Although observed intolerance has been reported among this population, higher levels of exercise and habitual physical activity have been shown to be important for the health and quality of life (QOL) of patients with CF. Unfortunately, however, exercise and physical activity remain underutilized in the clinical management of the disease. Recent research has provided a new perspective on the pathophysiology of

exercise intolerance in individuals with CF and the specific benefits of exercise in terms of improved pulmonary function. These new data can provide patients, families, clinicians, and scientists the necessary rationale for the inclusion of exercise and physical activity in the daily management of CF to improve clinical outcomes and survival among patients with the disease.

Significant challenges exist with respect to the use of exercise and physical activity as therapy for chronic diseases, particularly CF. Boas[3] reported that parents of children with CF perceive fewer benefits from exercise and greater barriers to physical activity than do parents of healthy children, with this view being more prevalent among parents of girls than of boys. In addition, the author found that fewer than half of the parents of children with CF knew that exercise performance was related to long-term prognosis or that exercise could be beneficial even among those with the most severe forms of CF. Further, in patients with CF, such issues as fatigue and the time required for treatments (ie, drug regimens and physiotherapy) make incorporating exercise into daily life even more challenging.

In addition to the challenges faced by patients with CF and their families regarding exercise as therapy, the medical and scientific communities have been engaged in a "chicken vs egg" type of debate. Simply stated, uncertainty exists as to whether exercise intolerance in patients with CF is primary or secondary to the disease. More specifically, the debate centers on whether exercise intolerance is due to impaired oxygen delivery or to intrinsic abnormalities in muscle function itself. Lamhonwah and colleagues (reviewed in this issue) have recently provided several lines of evidence demonstrating that the CF transmembrane conductance regulator (CFTR) is expressed in human skeletal muscle, and that CFTR dysfunction may contribute to the observed skeletal muscle weakness and exercise intolerance in patients with CF. These mechanistic observations are a crucial advance, as CF can now be considered a muscle disease, in addition to its previously defined effects on epithelial cells in the lung, pancreas, and digestive systems.

To build on the mechanistic observations that CF disease has a primary impact on skeletal muscle function and that skeletal muscle function is impaired in children with CF, it is important to recognize the established link between exercise capacity and lung function. Researchers have shown that a relationship exists between aerobic capacity, as assessed by traditional cardiopulmonary exercise testing, and survival in patients with CF.[4] Additionally, exercise training programs are effective in improving both exercise capacity and, in some cases, lung function in patients with CF. Interestingly, the positive benefits of exercise training programs appear to have long-lasting effects and are not limited to just aerobic training. Anaerobic and strength training programs have been shown to be effective as well. It is critical that the scientific and medical communities work toward developing clear exercise recommendations for patients with CF and that these recommendations are implemented as part of routine clinical practice.

Exercise training programs, although effective, may not always be easy to implement, supervise, and sustain. The current trend in health promotion is to encourage a lifestyle change of increased habitual physical activity, which refers to the level of activity incorporated into a person's daily life. Changes in habitual physical activity represent a lifestyle modification that results in long-lasting benefits. Habitual physical activity presents an interesting possibility for patients with CF to help incorporate daily exercise into their lives, thus improving their health and QOL. Evidence suggests that habitual physical activity is positively associated with lung function (forced expiratory volume in 1 second; FEV_1).[5] Schneiderman and coworkers reported a positive relationship between habitual physical activity and FEV_1 in girls when studied over a 2-year period. This study is ongoing, and a recent update over a longer evaluation period has confirmed the relationship between FEV_1 and habitual physical activity, also demonstrating that the relationship exists for males as well as for females.[6]

In summary, regular exercise and habitual physical activity are important for a patient with CF. Research has demonstrated the benefits of aerobic, anaerobic, and strength exercise training programs for health and QOL; however, patients with CF are faced with unique barriers and challenges to participation. In a recent review, Wilkes and co-workers have summarized research that has shown that increased levels of habitual physical activity slow lung function decline in patients with CF and that regular participation in a variety of activities may result in greater adherence in the long term.[7] Research is now available to justify the incorporation of exercise into the routine care of patients with CF. Education of

health care providers on the importance of exercise and habitual physical activity for patients with CF is needed, in order for exercise and physical activity to become key components of clinical practice and of the daily lives of patients with CF.

Commentary References

1. Corey M. Modelling survival in cystic fibrosis. *Thorax.* 2001;56(10):743.
2. Wilkes DL, Schneiderman JE, Nguyen T, et al. Exercise and physical activity in children with cystic fibrosis. *Paediatr Respir Rev.* 2009;10(3):105-109.
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4. Nixon PA, Orenstein DM, Kelsey SF, Doershuk CF. The prognostic value of exercise testing in patients with cystic fibrosis. *N Engl J Med.* 1992 Dec 17;327(25):1785-1788.
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CYSTIC FIBROSIS TRANSMEMBRANE REGULATOR EXPRESSION IN HUMAN SKELETAL MUSCLE

Lamhonwah AM, Bear CE, Huan LJ, Kim Chiaw P, Ackerley CA, Tein I. **Cystic fibrosis transmembrane conductance regulator in human muscle: dysfunction causes abnormal metabolic recovery in exercise.** *Ann Neurol.* 2010;67(6):802-808.

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The objective of the current study by Lamhonwah and associates was to examine cystic fibrosis transmembrane conductance regulator (CFTR) expression in human skeletal muscle. Previous research has shown that patients with CF experience exercise intolerance and skeletal muscle weakness, although the pathophysiology of these observations was poorly understood. Western blot analysis, confocal microscopy, and reverse transcription polymerase chain reaction (RT-PCR) were used to determine if CFTR is expressed in human skeletal muscle. Localization of CFTR in skeletal muscle was performed by immunoelectron microscopy.

The principal discovery by the authors is that CFTR is present in human skeletal muscle and that dysfunctional CFTR in human skeletal muscle has measurable pathophysiology. Specifically, the authors studied CFTR expression in human skeletal muscle by Western blot with anti-*CFTR* antibody L12B4, and demonstrated a single band with a molecular weight of 168 kDa. They then isolated the cDNA by RT-PCR and directly sequenced a 975bp segment that was identical to the human *CFTR* sequence. The cellular distribution of CFTR was localized to the sarcoplasmic reticulum, sarcotubular network, and sarcolemma. The investigators demonstrated staining of CFTR in the sarcolemma and sarcoplasm with the use of immunofluorescence microscopy. In a final set of experiments, the authors reported that they were able to activate *CFTR* chloride channels of wild-type isolated muscle fibers but were unable to do so in mutant F508del-*CFTR* muscle. These data in aggregate suggest a functional role of CFTR in the regulation of human skeletal muscle contraction.

The results of this study provide evidence for the expression of functional *CFTR* chloride channels in human skeletal muscle, with strong expression in the sarcotubular network. The authors speculate that *CFTR* dysfunction in skeletal muscle interferes with calcium regulatory channels or calcium adenosine triphosphatases (ATPases), which may contribute to the skeletal muscle weakness and exercise intolerance reported among patients with CF. Most importantly, the studies reported in this paper provide a mechanistic and metabolic rationale

for the observed exercise intolerance in patients with CF, and the fact that skeletal muscle dysfunction is primary and intrinsic to CF disease.

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METABOLIC PATHOPHYSIOLOGY OF SKELETAL MUSCLE DYSFUNCTION IN ADOLESCENTS WITH CYSTIC FIBROSIS

Wells GD, Wilkes DL, Schneiderman JE, et al. **Skeletal muscle metabolism in cystic fibrosis and primary ciliary dyskinesia.** *Pediatr Res.* 2011;69(1):40-45.

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The objective of this study was to identify and quantify muscle metabolic abnormalities in patients with CF compared with a respiratory disease control group with similar patterns of inflammation, infection, and bronchiectasis (primary ciliary dyskinesia [PCD]) and matched healthy control (HC) participants. The authors sought to differentiate the specific impact of CF on muscle metabolism from inflammatory factors associated with lung disease in general. The researchers used ^{31} phosphorus-magnetic resonance spectroscopy (^{31}P-MRS) to assess muscle metabolism *in vivo*, and to evaluate the function of creatine kinase, oxidative phosphorylation, and anaerobic glycolysis pathways during exercise and recovery. The use of ^{31}P-MRS, in conjunction with specifically designed exercise protocols, allowed for the noninvasive analysis of skeletal muscle metabolism in patients with CF.

The results suggested abnormalities in muscle bioenergetics both during rest (lower resting adenosine triphosphate [ATP] concentrations) and during short bouts of high-intensity exercise (higher end-exercise pH values) in patients with CF that are not observed in respiratory disease controls and in matched HCs. Since ATP is the primary energy source for muscle contraction, lower resting ATP levels might be indicative of a lower energy reserve available for exercise. Alternatively, low resting ATP levels might mean that ATP is being used at a higher rate at rest in patients with CF compared with HCs. Higher end-exercise pH could be interpreted as an impaired glycolytic capacity in the muscle. Both CF and PCD patients demonstrated delayed postexercise phosphocreatine recovery times compared with HCs, suggesting a nonspecific effect of respiratory disease on mitochondrial function.

In this research, the authors discuss the mechanistic pathophysiology of CF as it relates to skeletal muscle function *in vivo*. They described differences in resting energy levels, as well as in the metabolic pathways that provide energy during exercise. It appears as if some of these differences may be primary to the CF disease process itself. This is the first research paper that describes the intramuscular mechanisms of exercise intolerance in patients with CF. The results provide a rationale for the use of exercise therapies in this population, as physical activity and exercise have been shown to improve resting energy stores, as well as aerobic and anaerobic energy metabolism, in both healthy individuals and in patients with CF.

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FEV_1 AND VO_{2PEAK} AS PREDICTORS OF MORTALITY IN PATIENTS WITH CYSTIC FIBROSIS

Pianosi P, Leblanc J, Almudevar A. **Peak oxygen uptake and mortality in children with cystic fibrosis.** *Thorax.* 2005;60(1):50-54.

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Much of what is known about exercise in children with CF has come from cross-sectional studies that have shown that low FEV_1 and reduced lean body mass are correlated with lower peak oxygen uptake (VO_{2peak}) during incremental exercise testing. Previous research has shown that higher VO_{2peak} is significantly correlated with survival. According to the authors,

however, a dearth of longitudinal data are available on the assessment of VO_{2peak} in children. Pianosi and colleagues claim that longitudinal tracking of VO_{2peak} might be of interest, as it may offer some insight into understanding the relationship between fitness and survival in patients with CF. In the current study, the authors sought to analyze VO_{2peak} data and mortality over a 10-year period to determine if VO_{2peak} is a good predictor of mortality and, if so, if the rate of change in VO_{2peak} or the VO_{2peak} level is the more important variable. To this end, the research team performed annual spirometry per standard protocols, as well as exercise testing and tracking of disease outcomes (mortality or lung transplantation). Lung transplantation was interpreted as an endpoint, as the patients would not have survived without the procedure. VO_{2peak} was determined using a standard incremental step exercise protocol. Statistical analyses were performed to evaluate the influence of VO_{2peak} and FEV_1 on mortality.

Pulmonary function in the study population ranged from normal to severe airway obstruction. The overall decline in FEV_1 during the study period averaged 2.5% predicted. Similarly, the statistical model demonstrated that VO_{2peak} decreased 0.17 mL/min/kg per month. In both cases, the analysis showed that the rate of decline was more rapid in children who were older. For FEV_1, either a low initial FEV_1 with little change thereafter or a high FEV_1 with a significant rate of decline were both associated with mortality. The same pattern was apparent for VO_{2peak}.

The longitudinal analysis reported herein confirmed earlier findings that VO_{2peak} is a prognostic indicator of outcome in children with CF.[1] In the current research, patients in the lower tertile had a mortality rate of 60% in the subsequent 8 years. The statistical analysis showed that the effects of FEV_1 and VO_{2peak} on survival were of similar magnitude, although consideration of initial values and rates of decline supported the superiority of FEV_1 as a prognostic indicator. This study provides clear evidence for the value of physical fitness, participation in physical activities and regular exercise, in patients with CF. Further, establishing a foundation of aerobic fitness as early as possible may also be important, as the prognostic value of VO_{2peak} became more apparent when a patient's rate of lung function decline increased. The authors even hypothesized that patients with similar degrees of lung disease, as estimated by FEV_1, have different prospects for survival depending on their VO_{2peak}. Exercise testing delivers a comprehensive functional assessment of cardiovascular capacity, and is influenced by age, lean body mass, gender, genetics, and subjective effort. As such, exercise assessments may provide the clinician with valuable information on the overall health and function of their patients, and can be considered in light of specific pulmonary assessments as part of the clinical monitoring of patients with CF.

1. Nixon PA, Orenstein DM, Kelsey SF, Doershuk CF. The prognostic value of exercise testing in patients with cystic fibrosis. *N Engl J Med.* 1992 Dec 17;327(25):1785-1788.

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LONG-TERM EFFECTS OF TRAINING PROGRAMS FOR PATIENTS WITH CYSTIC FIBROSIS

Hebestreit H, Kieser S, Junge S, et al. **Long-term effects of a partially supervised conditioning programme in cystic fibrosis.** *Eur Respir J.* 2010;35(3):578-583.

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Studies investigating the effects of exercise programs in children and adults with CF have shown an increase in exercise capacity, a sustainment or increase in lung function, and enhanced QOL. Supervised and home-based programs have been studied, with both proving effective in improving the exercise capacity and health status of the participants. The primary objective of this study was to determine the effects of a home-based, partially supervised conditioning program on exercise capacity 12 to 18 months after the intervention program had ended. The variables of interest included VO_{2peak}, maximal work rate, physical activity levels

(assessed by accelerometry), pulmonary function (spirometry), and QOL (questionnaire). Participants were assessed at 3, 6, 12, 18, and 24 months. The participants in the intervention group were counseled and supported to increase their exercise and physical activity by a minimum of 3 x 60 minutes per week for the first 6 months of the study. Participants elected to engage in endurance-type sports, ball games, and/or strength training exercises, per personal preference. After the first 6 months, participants were counseled to maintain or increase their physical activity. Those in the control group were asked to maintain their current activity levels for the same period of time (12 months). During the second year of the study, all participants were free to change their physical activity behavior as they chose.

The intervention group increased their physical activity participation by 2.16 hours per week, of which about 1 hour was vigorous exercise. Participants in the intervention group had an increase in VO_{2peak} at 6 months, as well as during the open follow-up period at 12 to 18 months. Throughout the study, physical activity levels in the intervention group were significantly higher than in the control group. For pulmonary function, only forced vital capacity (FVC) was significantly higher (6%) in the intervention group compared with the control group ($P<.05$).

The primary finding of this study was that the positive health effects of a 6-month, home-based, partially supervised conditioning program can be observed for up to 12 months or longer after the end of the intervention. Beneficial effects were noted in terms of exercise capacity, physical activity participation, and FVC, all of which are relevant and important determinants of health in patients with CF.

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ANAEROBIC TRAINING IN CHILDREN WITH CYSTIC FIBROSIS

Klijn PH, Oudshoorn A, van der Ent CK, van der Net J, Kimpen JL, Helders PJ. **Effects of anaerobic training in children with cystic fibrosis: a randomized controlled study.** *Chest.* 2004;125(4):1299-1305.

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Regular aerobic exercise has positive effects on the aerobic capacity of patients with CF, and has also been associated with prolonged survival and enhanced QOL. To date, limited research has focused on the effects of anaerobic fitness in the CF population. This is disappointing, as children's natural pattern of play is characterized by short bursts of intense activity interspersed with periods of low-intensity exercise or even rest. Therefore, anaerobic training programs may be highly suitable for children with CF. Further, children with CF have been shown to have reduced anaerobic performance. The aim of this research was to investigate the effects of an anaerobic training program on exercise performance, lung function, and QOL in children with CF.

Participants were assessed at baseline, within 7 days after completing a 12-week supervised training program, and once again after a 12-week period in which the children were able to participate in physical activity as they wished. Measurements included anthropometry, pulmonary function, anaerobic exercise performance using the traditional 30-second Wingate cycling test, and aerobic performance using a standard incremental ramp protocol. The anaerobic training program consisted of a 30- to 45-minute training session that was performed 2 times per week for 12 weeks (the program can be downloaded at http://www.chestjournal.org/cgi/content/full/125/4/1299/DC1).

Perhaps the most interesting result of this study was the fact that adherence to the training program was 98%. This is an excellent rate for a training intervention study. Pulmonary function was unchanged in both the intervention and control groups, although the training group demonstrated significant improvements in both aerobic (+5.7% predicted, $P<0.05$) and absolute anaerobic (+11% to +12%, $P<0.001$) exercise performance. QOL in the physical functioning domain, as assessed by questionnaire, was improved in the training group only. Anaerobic performance and markers of QOL, but not aerobic fitness, remained elevated during the follow-up period.

These results demonstrate that children with CF can improve their aerobic and anaerobic exercise capabilities, as well as their perception of physical functioning, in as few as two 30-minute anaerobic training sessions per week. The authors suggest that anaerobic training programs may have higher rates of participation and compliance, as anaerobic exercise is similar to the nature of children's play. Interestingly, anaerobic function and perceived QOL remained elevated up to 12 weeks after completion of the formal training program. It is also interesting to note that the anaerobic training program had a positive effect on aerobic capacity as well. Unfortunately, an improvement in pulmonary function was not observed in the training group. The direct effects of training programs on pulmonary function in patients with CF remain equivocal, with some studies demonstrating positive effects and others showing no improvement. To date, no studies have demonstrated adverse effects from exercise on pulmonary function in patients with mild to moderate CF.

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