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Does exercise improve lung function in a young adult with Cystic Fibrosis?

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The University of Akron 2021

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Abstract

Cystic fibrosis (CF), is an inherited genetic disease that leads to damage of the respiratory system. This disease not only impacts the lungs, but most organ systems of the body. Normal fluids in the body become thick, clogging important pathways in the body such as in the lungs and the pancreas. PURPOSE: To examine the effect of cardiorespiratory exercise training on lung function. The purpose of this study was to examine how introducing an aerobic-based exercise training program impacts patients with CF. The question is, "Does exercise improve lung function in a young adult with Cystic Fibrosis?". METHODS: A 22-year-old female volunteer participant agreed to complete a 7week exercise training program, consisting of three days of cardiorespiratory training and one day of active recovery. These sessions lasted from 30-45 minutes in duration and were programmed at a light to moderate intensity. Pre and post exercise testing was completed by the participant before and after this exercise program. Forced expiratory volume (FEV1) was recorded once before, during, and after completing the exercise program. The Rockport walking test protocol was used to evaluate cardiorespiratory endurance while the curl up test was used to evaluate muscular endurance. RESULTS: The times taken to walk a mile, using the Rockport Walk Test protocol were 16:03 minutes and 14:57 minutes, respectively. The results from the curl up test before and after were 34 and 47, respectively. Finally, the values for FEV1 were measured to be 83 and 89, respectively. DISCUSSION: Due to IRB Covid-19 protocol, this case study was done remotely. It is possible that if exercise sessions or testing did not have to be the sole responsibility of the participant to complete, they would have achieved more successful performance. Also, it should be noted that this program lasted seven weeks. With more time, a greater improvement in fitness may have been achieved. Still, there were improvements in the three variables measured.

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Introduction

Cystic fibrosis (CF) is caused by a mutation of the CF gene on chromosome 7, and is associated with reduced airway function, low exercise capacity, and pulmonary infection (Van Iterson et al., 2016). It is not uncommon for individuals diagnosed with this disease to spend at least one and a half hours per day participating in treatments such as airway clearance strategies or taking antibiotics and other medications (Dwyer et al., 2020). Regular exercise is associated with increased cardiorespiratory fitness, pulmonary function, and improved quality of life (QOL) (van de Weert-van Leeuwen et al., 2013). Only 21-57% of adults with CF report participating in regular exercise programs (Dwyer et al., 2020). In addition, "exercise has been described to affect disease incidence by modulating the immune system" (van de Weert-van Leeuwen et al., 2013).

Given that exercise provides an abundance of benefits for all adults, this study specifically examined how an exercise intervention program can impact the pulmonary function of a young adult with cystic fibrosis. This case study examined cardiorespiratory exercise and how beginning a regular exercise routine this may improve lung capacity in a young adult female with CF. It provides insight to possible future and current treatment options and builds on the importance of using exercise as medicine. Encouraging regular cardiovascular activity in more young adults with CF may allow exercise to help manage this disease.

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Literature Review

What is Cystic Fibrosis

Cystic fibrosis is an extremely complex disease. This disease is caused by a mutation of the cystic fibrosis (CF) gene on chromosome 7, leading to "…improper transport of CF transmembrane conductance regulator (CFTR) within lung tissue at the epithelial cell level" (Van Iterson et al., 2016). CFTR plays a crucial role in airflow, gas transfer, and abnormal ventilation (Van Iterson et al., 2016). The CFTR gene encodes an ATP regulated ion channel that is revealed in many tissues (van de Weert-van Leeuwen et al., 2013). Along with chronic pulmonary infections and inflammation, CF patients struggle with exercise intolerance, muscle cachexia, and inflammation (van de Weert-van Leeuwen et al., 2013). The inflammation caused by CF leads to progressive pulmonary disease as well (van de Weert-van Leeuwen et al., 2013).

Peripheral vascular function is likely in CF patients, however more research on this exact pathophysiology is warranted (Van Iterson et al., 2016). This genetic complication impacts every aspect of an individual's life. CF is a disease of multiple systems and can often lead to serious complications. Infections such as Staphylococcus aureus, Haemophilus influenzae, and Pseudomonas aeruginosa, especially when occurring early in life, can lead to chronic infection and worsening of the respiratory system. (Bradley et al., 2006). A primary symptom of CF is low exercise capacity, especially in regard to ventilation and gas-exchange (Van Iterson et al., 2016). This is due to the buildup of sputum, causing breathlessness. Many interventions are done to combat this. Examples of these are airway clearance and physical training (Bradley et al., 2006).

There is an emotional impact of CF as well. Children and adolescents have reported feeling physically or emotionally vulnerable, resentment, loss of independence or opportunities, isolation, and disempowerment" (Ohn & Fitzgerald 2018). As mentioned, this disease is associated with a declining pulmonary function. Pulmonary infection and impaired lung function are main causes of morbidity and mortality in these patients (Tucker et al., 2017). Many factors inhibit exercise capacity such as impaired oxygen uptake, oxidative stress, and abnormal pulmonary function. (Tucker et al., 2017). One of the potential modifiable factors that may improve CF include exercise, as well as avoiding smoke exposure, household income, and health insurance (Collaco et al., 2014).

Treatments for Cystic Fibrosis

While the amount of effective therapies are increasing, there is no cure for CF. According to Ohn & Fitzgerald (2018), "The price for an improved life span is the commitment of adherence to a time-consuming, repetitive, labour intensive and often complicated daily treatment regimen." Therefore, adherence to treatment is vital for management. Daily therapy and treatments can last over an hour, using standard techniques for treating cystic fibrosis such as airway clearance, pancreatic enzyme replacements, oral antibiotics, and inhaled mucoactive medication. (Dwyer et al., 2020). Physical therapy regimens are part of a comprehensive plan. The systematic review written by Bradley et al. (2006) demonstrated that short term studies (up to one month) have both shown increase in lower limb strength due to aerobic activity and significant improvement in exercise tolerance. The study stated:

> "In two short-term trials, no significant differences were found in spirometry after aerobic training compared with bronchial hygiene or no specific physical training. Longer

> term trials showed that aerobic physical training had no significant effect on FEV1 but a significant and positive effect on FVC compared with normal activity." (p.197)

These long-term studies were up to three years in length. It can be said that there is poor adherence to exercise, at a value around 40-50%. A goal of future research is to improve this. It should be noted that single, short- and long-term studies looking at exercise have shown benefits to the disease status and life of CF patients. There is also no evidence to support or refute the substitution of airway clearance sessions with physical training. It was published that there was not enough evidence to support or refute the practice of exercise. In fact, both anaerobic and aerobic training programs are important to these patients. Physical training is already part of the care package offered to most patients with CF, and there is no evidence to actively discourage it (Bradley et al., 2006). The studies included in this review warrant future research in the area of exercise and management of Cystic Fibrosis.

Outside of medication and airway clearance, patients participate in physiotherapy and exercises, inhaled antibiotics and mucolytics, a high-fat, high-calorie diet and extra nutritional supplements, and pancreatic enzyme replacement therapy (Ohn & Fitzgerald 2018). The Cystic Fibrosis Foundation (2020) states that people with CF have an estimated energy requirement of 1.5 to 2 times higher than those without the disease. This is because more energy is required to breathe normally. As they are often fighting infections in the lungs, digestion is compensated. A dietician will work with people who have CF to add calories to the food they already eat, as gaining weight is often a goal in children and teens. Data shows that improvements in nutrition is associated with improvements in pulmonary function (Cystic Fibrosis Foundation, 2020). A lack of commitment and or compliance with CF treatment can result in a more rapid progression of

the disease. Treatment for cystic fibrosis is multifaceted and individualized to meet patient needs. Therefore, a multi-disciplinary CF team is necessary to troubleshoot and overcome barriers in many different areas (Ohn & Fitzgerald 2018). These patients are encouraged to exercise in order to maintain not only fitness but an overall better quality of life. (van de Weert-van Leeuwen et al., 2013).

Benefits of exercise

Exercise training improves muscle strength, overall exercise capacity, and reduces breathlessness. Tucker et al. (2017) suggests that exercise capacity can also predict mortality. These authors state, "Preserving both exercise capacity and pulmonary function have significant implications for overall health and wellbeing in patients with CF (p.752)." Further research can support the beneficial relationship between chronic exercise training, pulmonary function, and airway inflammation. An increase in spirometry function is an acute exercise response in CF patients, likely due to shear stress in the airways and subsequent bronchodilation. Overall, lung function appears to improve after exercise training for patients with CF. The specific parameters for a treatment response such as modality, duration, intensity, and frequency are still unknown. While this was an acute study, the findings show that a small volume of exercise can transiently improve lung function and are promising to be used as a foundation in acute exercise studies to gain more knowledge of pulmonary function (Tucker et al., 2017). Another study by Dwyer et al. (2020), suggests that that exercise training is recommended for patients with CF to improve their quality of life as well as lung function. In a systematic review assessing exercise in healthy and patient populations, the theme that exercise improves physical and psychological health was

identified in 100% of the studies included. This suggests that since exercise is beneficial to all young adults, it may specifically be beneficial to CF patients.

According to Collaco et al., (2014) "A recent larger prospective study of exercise in children with cystic fibrosis (n = 212) did see an association between increased habitual physical activity and a reduced decline in FEV1" (p.2). Therefore, these results suggests that exercise patterns in childhood may carry over to adulthood. van de Weert-van Leeuwen et al., (2013) discuss the role of exercise in lung function as well. Exercise is associated with higher baseline lung function and a less rapid decline in lung function. Another positive effect of moderate and regular exercise is anti-inflammatory responses, as well as reduced disease incidence, and viral infection susceptibility. Chronic exercise response limits inflammatory response in conditions such as obesity, chronic heart failure, atherosclerosis, and diabetes. Therefore, exercise could benefit chronic inflammatory and infectious diseases such as cystic fibrosis (CF). Exercise is also an important part of muscular strength, cardiorespiratory fitness, and quality of life for CF patients. Finally, exercise modulates the immune system; however, the underlying mechanisms of this concept have not been deeply studied in CF patients (van de Weert-van Leeuwen et al., 2013). It can be said that exercise is beneficial because it leads to a greater stretch and recoil of inspiratory muscles and possibly an increase in mucous clearance (Tucker et al., 2017).

Exercise and Cystic Fibrosis Patients

Many individuals with CF also have exocrine pancreatic insufficiency, which leads to poor weight gain and the concern of increased caloric expenditure with exercise (Collaco et al., 2017). This should be taken into consideration regarding CF and exercise. There are CF-specific advantages, disadvantages, facilitators, and barriers to exercise regarding pulmonary function

that are of relevance to CF patients. (Dwyer et al., 2020). Participants of an elicitation interview study conducted by Dwyer, Griffin, Bye & Allison (2020) were asked a series of open-ended questions about CF patients and exercise. Those with CF were asked to answer regarding their own beliefs, while relatives or friends were asked about their friend or family member with CF, regarding exercise. Ultimately, the information collected from patients with CF were used, as it was found that friends and relatives provided similar information. The results are summarized as follows:

> A few advantages of exercise include "keeping fit and healthy (68%); improve lung function and decrease breathlessness (56%) and decrease cough and clear the lungs (44%)…feel better and happier (60%); feel stronger and have more energy (28%); improve body image (24%) and be fun or social (20%). Also, 16% of participants stated an advantage of exercise was to reduce the time required for airway clearance techniques." Disadvantages included "feel breathless (36%); cause injury (24%) and cough (16%)…feel tired or reduce energy (16%); not enjoy exercise (16%); feel embarrassed to exercise in public (12%) and feel frustration when unable to keep up with others (12%). Also, 24% of participants commented that a disadvantage of exercise was to be time-consuming." (pp. 288-289)

Complications to consider when choosing any type of physical therapy (including airway clearance as well as physical training) for CF include dyspnea, gastro-esophageal reflux, bronchospasm, hypoxemia, pain, fatigue, dehydration, and hemoptysis, and to a lesser extent, rib fractures and pneumothorax. However, the latter conditions are rare (Bradley et al., 2006). Exercise improvement also depends on more than one factor such as frequency, type of training,

and exercise intensity. Tucker et al. (2017) explained that there is no guarantee pulmonary function will improve in patients even with exercise training, and it is likely that a higher intensity of exercise may lead to greater improvements in pulmonary function. The exercise principle of individuality should also be noted as someone with a less advanced fitness level has more opportunity to improve. Also, there have been significant decreases in the lung clearance index after a single exercise session, likely due to dynamic hyperinflation of distal airways during exercise or the sputum clearance. The authors continued that the increase in pulmonary function is likely related to an acute reduction in lung inflammation. Also, the participation in exercise and not specifically how intense that exercise is, influences changes in the lung clearance (Tucker et al., 2017). Since fitness levels are reduced in those with chronic illness, any exercise program needs to account for previous exercise experience, relative level of fitness, and tailored to intensity, intervals, volume, and timing (van de Weert-van Leeuwen et al., 2013). The inflammatory response might be different in different subjects as well. Therefore, if inflammation is aggravated, exercise may need to be avoided during periods of active symptoms. (van de Weert-van Leeuwen et al., 2013). Exercise adherence is necessary for training benefits and advancements. Therefore, it is reasonable to conclude that exercise habits formed during childhood could lead to healthy lifestyle habits in adulthood.

Methodology

This individual case study was approved by the author's university Institutional Review Board. This case study methodology was chosen because exercise needs to be specific to individual needs. A volunteer, 22-year-old adult with Cystic Fibrosis agreed to participate in this case study. She was given an informed consent form to complete and instructed to obtain

medical clearance, as this participant did not regularly participate in exercise prior to the study. The Informed Consent is included as Appendix A. The participant was given written instructions to complete self-administered pre and post exercise testing, due to IRB COVID-19 protocols. She was instructed to complete the Rockport 1-mile walk test and the Curl up test, recording these results with her initial lung function, as measured by spirometry. The curl up test involved the participant lying in a supine position, with their knees bent at 90 degrees and feet flat on the floor. To the beat of a metronome, the participant lifted their shoulders off of the ground, reaching toward a piece of tape past her fingertips (Pescatello et al. 2014). The Rockport test protocol includes walking a one mile as fast as possible, while not breaking into a run. The time for this is recorded as well as heart rate after walking (Swain et al. 2014). These tests represented the variables of cardiorespiratory endurance, muscular endurance, and lung function, respectively. Lung function was recorded by spirometry using the ZEPHYRx LLC app, Breathe Easy (v1.9.4). Lung function is assessed as part of the daily treatment regimen for this individual but was specifically noted at three points in the study. The participant was also provided with warning signs of negative physiological responses to acute exercise in the case of an emergency. Instructions for testing and exercise are included in Appendix A, B & E.

After recording test results, the participant was given a remote exercise program to complete from home. This program lasted seven weeks. It consisted of a variety of free, online workouts, that were previewed for effectiveness, duration, and intensity. Most of the workout videos required minimal space and equipment, making it easy for the participant to complete. The workouts ranged from 20-45 minutes. They were programmed to start at a light intensity and got progressively more difficult. Each day included a 5-7 minute warm up and cool down. Three days of primarily cardiorespiratory workout routines were completed with one day of active

recovery. The variety of exercise modalities gave the participant exposure to different types of cardiorespiratory activity. Different modalities included dance cardio, low intensity interval training, kickboxing, and bodyweight training. With each session, modifications were included if needed. This gave the participant autonomy to modify or advance as desired. Instructions were given to the participant to explain exercises. These were in video and written format. This encouraged proper form and safety when exercising, allowing the participant to be educated. The participant was given the freedom to withdraw from the program at any time. Each weekly workout plan was compiled into a single document and sent via email to the participant at the beginning of each week. A sample exercise plan is included in Appendix C. Communication occurred with the participant between each week of the exercise program to assess preferences, individual needs, and rate of progression. This allowed the program to advance in the most effective way possible. Half-way through the program, a questionnaire was given to the participant to assess compliance (Appendix D). After completing seven weeks of exercise, the same exercise tests that were done at the beginning of the program were completed. Subjective interview questions (Appendix F) were completed at the end to consider the patient's perspective and mindset. Results from the pre and post exercise testing were compared to evaluate improvement.

Results

Two exercise tests were used for this study. The first was the Rockport Walk test (Swain et al. 2014). The second was the Curl up muscular endurance test (Pescatello et al. 2014). The participant's FEV1 was also recorded using the ZEPHYRx LLC app, Breathe Easy (v1.9.4). These three values were recorded before the first week of exercise, and after the seventh week of

exercise. The data for before and after was examined for relative differences. Table 1 shows pre and post assessment results. Relative pre and post assessments are summarized in Table 2.

Table 1

Pre and Post Assessment Results

Test Type	Before	After 7 Weeks
Rockport Walk Test	16:03 minutes	14:57 minutes
Curl Up Test	34 curl ups (Average)	47 curl ups (Well above average)
FEV1	83%	89%

After completing the seven (7) week exercise program, the participant showed improvements in cardiorespiratory, muscle endurance, and lung function tests. The participant improved her 1-mile walk completion time by slightly more than a minute. Since she did not take her heart rate, a rating could not be determined. She was able to complete thirteen (13) more curl ups, and her FEV1 score improved by 6.7%. The ratings for the test were determined using Pescatello et al. (2014). These differences are summarized in Table 2.

Table 2

Summary of Differences Pre to Post Assessments

Test Type	Rockport	Curl up	FEV1
Difference in value	1:06 minutes	13 curl ups	6
Percent Difference	~6.9% higher	~28% higher	~6.7% higher
Conclusion	Improvement	Improvement	Improvement

Compliance was evaluated halfway through the program to determine if the participant wanted to continue exercise (Appendix D). After completing half of the workout program, the participant reported only having missed two workout sessions. She felt that she enjoyed the variety of exercise modalities and was challenged on average, to be working at an 8 on a 1-10 scale. The most prominent barrier was time to complete a full workout, including a warm-up and cool down. However, this was successful on most days.

By the end of the program, the participant reported "I can breathe easier and clearer- I can also feel that my stamina has increased because small things like folding laundry or walking up the stairs make me out of breath, and it has become easier to do things like that! She also stated that it was challenging to get started. She stated, "I knew it would be hard since my lungs are not used to that kind of cardio yet, but after a week or so it got easier… I felt I could notice a

clear difference in the time of this program! I was able to move more mucus out of my lungs and my stamina has increased so it made a big improvement in my opinion!"

Discussion

These findings provide insight to how exercise programming could impact CF treatment in the long term. Based on previous research, it was expected that cardiovascular exercise training can improve lung function in this specific patient population. This agrees with the literature previously discussed, in that exercise can show improvements in the health of CF patients. The literature did not provide specific parameters for frequency, intensity, and duration for fitness improvement. The results of this study suggest improvements can be noted with a moderate-level intensity program in as little as seven weeks.

This type of research design was influenced by the commitment of the individual. It required strict compliance and dedication to the exercise program This individual is a young adult living on her own. This is significant because she was fully autonomous in making lifestyle decisions. Without the continuous effort of choosing to exercise each week, there would not have been as much effort into improving her health. The commitment to making a behavior change led to positive choices. If her circumstances were different, for example if she was a child, or providing for someone else, she would have more likely been under the authority or influence of someone else.

The three variables measured, cardiorespiratory endurance, muscular endurance, and FEV1, all improved by the end of seven weeks. The time to complete the Rockport Walk test and the lung function improved by over 6%, while muscular endurance improved over 25%.

Overall, the participant stated that she was previously intimidated by exercise. Now, she feels that exercise can fun and she is stronger than she thought. She hopes to continue exercise.

For future research, a longer study may lead to greater improvements. Also, this test could have been done with more than one individual, allowing for statistical analysis to be done. Since there was essentially one value for each test result, running a paired sample t-test was not possible. Ideally, this would have allowed evaluation of any significant differences. In this study, percent improvement was used to determine progress instead. Also, due to Covid-19 IRB protocol, this study had to be done remotely. It was unsure if she would have consistent access to a fitness center or be able to exercise with other individuals. Therefore, exercise was completed in a home and an outdoor setting. Completing an exercise program with access to cardiovascular machines or other equipment could have also influenced results. The restrictions put in place to complete this study affected social interaction. If this study were to be repeated, it would be preferable to measure body composition, flexibility, and muscular strength as well. However, it was more realistic for this research design to study only three variables. The only aspect of the study that the participant did not comply with was measuring heart rate during the Rockport exercise test. Using heart rate in this test would provide more accurate results.

Designing a research project was a unique experience for me. The opportunity to develop my own research idea from the beginning was an important milestone in my academic career. The process of research design, as well as executing a case study, was valuable and furthered my education in many positive ways. I appreciated the autonomy, as well as the academic support, that I had in creating this study. Exercise and behavior change is something I am passionate about. I enjoyed being able to apply my undergraduate degree in a practical and hands on way. It

was more challenging than I expected. A few of the challenges included to adapting to schedule changes and relying on the compliance of the individual participating in this case study. I knew there was a significant possibility that this study would be inconclusive. However, I am so thankful for the participant that chose to complete this program in its entirety. I could not have asked for better participation and outcome. It is motivating to have worked on something that inspired behavior positive behavior change. Overall, this was a very positive experience and I am extremely grateful for all the participant's hard work.

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Appendix A: Informed Consent

Informed Consent Document

Title of Study: Does exercise improve lung function in a young adult with cystic fibrosis?

Introduction: You are invited to participate in a research project being conducted by Hanna Mackey, in the School of Sport Science and Wellness Education, at The University of Akron.

Purpose: This is a case study. This study will determine if an 8-week exercise intervention program will result in an improvement in your lung capacity.

Procedures: Medical clearance will be obtained due to ACSM's screening guidelines for exercise testing. You will complete a pre and post exercise assessment to assess cardiorespiratory fitness and muscular endurance. Cardiorespiratory endurance will be assessed using the Rockport Walk test. Muscular endurance will be assessed using a curl up test. You will then complete a 30-45-minute exercise routine 3-5 days per week, for eight consecutive weeks. It will encompass cardiovascular training, resistance training, and flexibility. You will complete the program in either your home, outdoors, or at a local fitness center. It will use minimal equipment and be designed to fit your fitness levels. The program will progress and get increasingly more difficult over the course of 8 weeks. The same exercise tests will be completed after the 8-weeks. Lung function will be assessed with at home Bluetooth treatment equipment throughout the exercise intervention.

Exclusion: If medical clearance is not obtained, you will not be able to participate.

Risks and Discomforts: There are several risks that are part of the decision to participate in any exercise program. These include musculoskeletal injury or the rare instance of sudden cardiac death. This can be viewed in detail according to ACSM’s guidelines. These risks are minimized with medical clearance and exercise testing to determine appropriate exercise intensity.

https://www.acsm.org/docs/default-source/publications-files/acsm-guidelines-download-10th-edabf32a97415a400e9b3be594a6cd7fbf.pdf?sfvrsn=aaa6d2b2_0

Does exercise improve lung function in a young adult with Cystic Fibrosis?

Benefits: The benefits to you for participating in this study may be improved fitness levels, such as cardiorespiratory or muscular endurance, flexibility, or improved respiratory and lung function. However, you may receive no benefit from participating in this study. Long term adherence to an exercise program is associated with reduced risk of chronic disease. The benefits can be further read in more detail using the provided link.

https://www.acsm.org/docs/default-source/publications-files/acsm-guidelines-download-10th-edabf32a97415a400e9b3be594a6cd7fbf.pdf?sfvrsn=aaa6d2b2_0

Right to refuse or withdraw: Participation in this study is voluntary. You have the right to refuse or withdraw participation at any time.

Anonymous and Confidential Data Collection: Any identifying information collected will be kept in a secure location and only the researchers will have access to the data. You will not be individually identified in any publication or presentation of the research results. Only aggregate data will be used. Your signed consent form will be kept separate from your data, and nobody will be able to link your responses to you.

Confidentiality of records: Your name will not be included in this study. The age range of young female adult, and the condition of Cystic Fibrosis will be the only personal information published in this study.

Who to contact with questions: If you have any questions about this study, you may contact Hanna Mackey at 330-636-6032 or Melissa Smith at mgsmith@uakron.edu. This project has been reviewed and approved by The University of Akron Institutional Review Board. If you have any questions about your rights as a research participant, you may call the IRB at (330) 972-7666.

Acceptance & signature: By signing this consent form, you affirm your understanding and willingness to be involved in the study.

I have read the information provided above and all of my questions have been answered. I voluntarily agree to participate in this study. I will receive a copy of this consent form for my information.

_____ _____

Participant Signature **Date**

Appendix B: Verbal Script

As you exercise, pay attention to your body. If you have any of the following symptoms, slow down or stop exercising right away. Call your doctor to discuss these symptoms as soon as possible. Call 911 immediately if they persist or increase in severity.

- Any chest pain (angina) -- including pain, tightness, or heaviness in the chest, neck, jaw, or arms
- Light-headedness or dizziness
- Nausea or vomiting
- Extreme fatigue (tiredness)
- Muscle pain
- Excessive sweat or cold sweat
- Vision problems
- Numbness or tingling in hands or feet
- Irregular or too-rapid heartbeat
- Unusual shortness of breath (more than you'd expect for your level of exercise or enough that you cannot carry on a short conversation)

Appendix C: Sample Exercise Plan

Week 1: 3 beginner workouts, light to moderate intensity

Check this out for proper squat form Check this out for proper knee push-ups

Warm up (do this each day)

Day 1: 15-minute beginner cardio workout

Check this out for proper squat form

Check this out for proper knee push-ups

Circuit 1 modifications to make it easier: *Rest between circuits if you need to*

- Step in place instead of jog
- Do not add jumps
- Slow down the pace at any time
- Step out instead of jumping jacks

Circuit 2-3 modifications (only changing last two exercises):

- Take out jumping jack
- Take out toe touches
- Only do a half lunge

Day 2: Full body beginner strength (20 minutes)

Most of these have modifications, but if you need them:

- Take out jumps/slow down pace
- Limit your range of motion
- Only do one arm then one leg (for bird dogs)
- Take a 30 second active rest between all exercises (not just the beginning)

Day 3: 15-minute beginner HIIT

- Don't jump at the top of your burpee
- Do not use weights
- Do push up on your knees!
- Take out the squat

Cool down

Recovery Day Yoga Routine

Appendix D: Compliance Check in

Week 4 Workout Check in

1) Do you feel you have time to complete this workout plan?
    a. Participant response: “Yes, I do feel that I have time to complete these!”
2) Have you missed any workouts? If so, how many?
    a. Participant response: “I missed 2- one in the first week and one recently, but I have been consistent other than that!”
3) Are you able to successfully complete the warm-up and cool down?
    a. Participant response: “Yes, and I like them! Especially the dancing one since it’s fun!”
4) Do you feel you have enough education to complete these workouts safely and effectively?
    a. Participant response: “Yes, Hanna gave me resources like YouTube links, or gave me modifications for the workouts so I felt I had enough education to do so.”
5) On a scale of 1-10, how hard are you typically working? As many times as you can, jot down this number during the week.
    a. Participant response: “I would say most days I am at an 8 or higher- but this is because of the constant movement in workouts and my lungs are not the greatest at that (exerting energy in that way).”
6) Are some days significantly easier than others?
    a. Participant response: “I don’t think I would say one is harder than the others, other than the rest day workout.”
7) What are your likes/dislikes so far, and how can I make this process better for you?
    a. Participant response: “I really enjoy the people and groups Hanna sends me! They make it fun, and they also help me as I go through the workouts, such as counting or encouragement. The only “dislike” even though it isn’t really a dislike, is the workouts that are longer than 35-40 minutes, only because I like to do the warm-ups and cool downs and then the workouts, so it takes longer, but I am still okay with doing them!”

Feel free to write down a sentence or two in your journal about noticing self-improvement, possible soreness, or pain. Or, any other relevant comments you have.

Appendix E: Self-administered Exercise Testing instructions

Self-administered testing instructions

Please read all instructions before completing the tests.

Self-Administered Curl Up Test

- Watch this video: https://www.youtube.com/watch?v=uLA_9kx1AkY
- View the following website: https://exrx.net/Testing/CurlUpTests
- Use this curl up cadence: https://www.youtube.com/watch?v=RW6OssVmJBI

- Lay on the mat with knees bent at 90° and feet on the floor. Your arms will touch a piece of masking tape. A second piece of tape will be beyond this piece.
- Set a metronome or timer at 40 beats per minute
- At the first beep, slowly lift your shoulder blades off the mat until fingertips reach the second piece of tape.
- At the next beep, slowly return your shoulder blades to mat by flattening lower back
- Repeat
- One repetition is counted each time shoulder blades touch the floor. Performs as many curl-ups as possible without stopping, up to a maximum of 75 repetitions.
- The test is terminated if the cadence is broken.

Does exercise improve lung function in a young adult with Cystic Fibrosis?

Self-Administered Rockport Walk Test

This test can be done on any flat surface that is one mile long (ex., UA Rec center, a park with mile markers, the street you live on). A heart rate monitor is preferred for this test. If you do not have one, use the following instructions to measure your own.

- View: https://www.whyiexercise.com/rockport-walking-test.html
- See other appendices for the Rockport test
- View this video (https://www.youtube.com/watch?v=K8ryHOgfTtY) and practice taking your radial pulse.

Begin test:

- Walk as quickly as possible for one mile. It's important to maintain a steady pace for best results.
- Record your time in minutes and seconds.
- Take your pulse immediately *after* completing the mile. (If you don't take your pulse often, practice before you take the Rockport walking test. With your palm facing up, place your fingers gently over the thumb side of your wrist and feel for your pulse.)

Record all results in your exercise journal. We will discuss these results over a virtual conference, and I will do the appropriate calculations.

Appendix F: End of program questions and participant responses

1) Do you feel that your cardiorespiratory fitness improved after completing this program? If so, describe how.

Participant response: “Yes! I can breathe easier and clearer- I can also feel that my stamina has increased because small things like folding laundry or walking up the stairs make me out of breath, and it has become easier to do things like that!”

2)Discuss your overall experience with this program. What was the most challenging? What did you enjoy or not enjoy? Do you feel it was long enough to notice improvement?

Participant response: “I absolutely loved getting to do this program! The most challenging part was honestly just starting. I knew it would be hard since my lungs are not used to that kind of cardio yet, but after a week or so it got easier! I enjoyed the variety of programs I was doing- some days were kickboxing and dancing, and some were abs, arms, and legs focused and I really liked that! I felt I could notice a clear difference in the time of this program! I was able to move more mucus out of my lungs and my stamina has increased so it made a big improvement in my opinion!”

3)Did this program change how you feel about or view exercise? Do you see exercise as something you will continue to participate in?

Participant response: “Yes! I think I get scared to work out because I know that I am not good at it, but this program made me realize that exercise can be fun, and I am stronger than I think! Yes, I will continue to participate in exercising!”

4)How do you think exercise can impact patients with CF? What role should exercise play in treatment? Has this program changed how you view exercise and CF?

Participant response: “I think that exercise can really make a difference in lung function, breathing, and just overall health in patients with CF. It helps us to get our lungs working harder and helps us build better lung functions that can lead to less infections, hospitalizations and more. I think that exercise should be a pretty big part of CF, although it can be hard to start if you haven't exercised before. I know CF patients who have lung functions of 20% and it would be very hard for them to even walk, let alone exercise. BUT I think it's important to talk about the importance of exercising ahead of time so that it doesn't get to that point! Seriously had SO much fun doing this ☺”