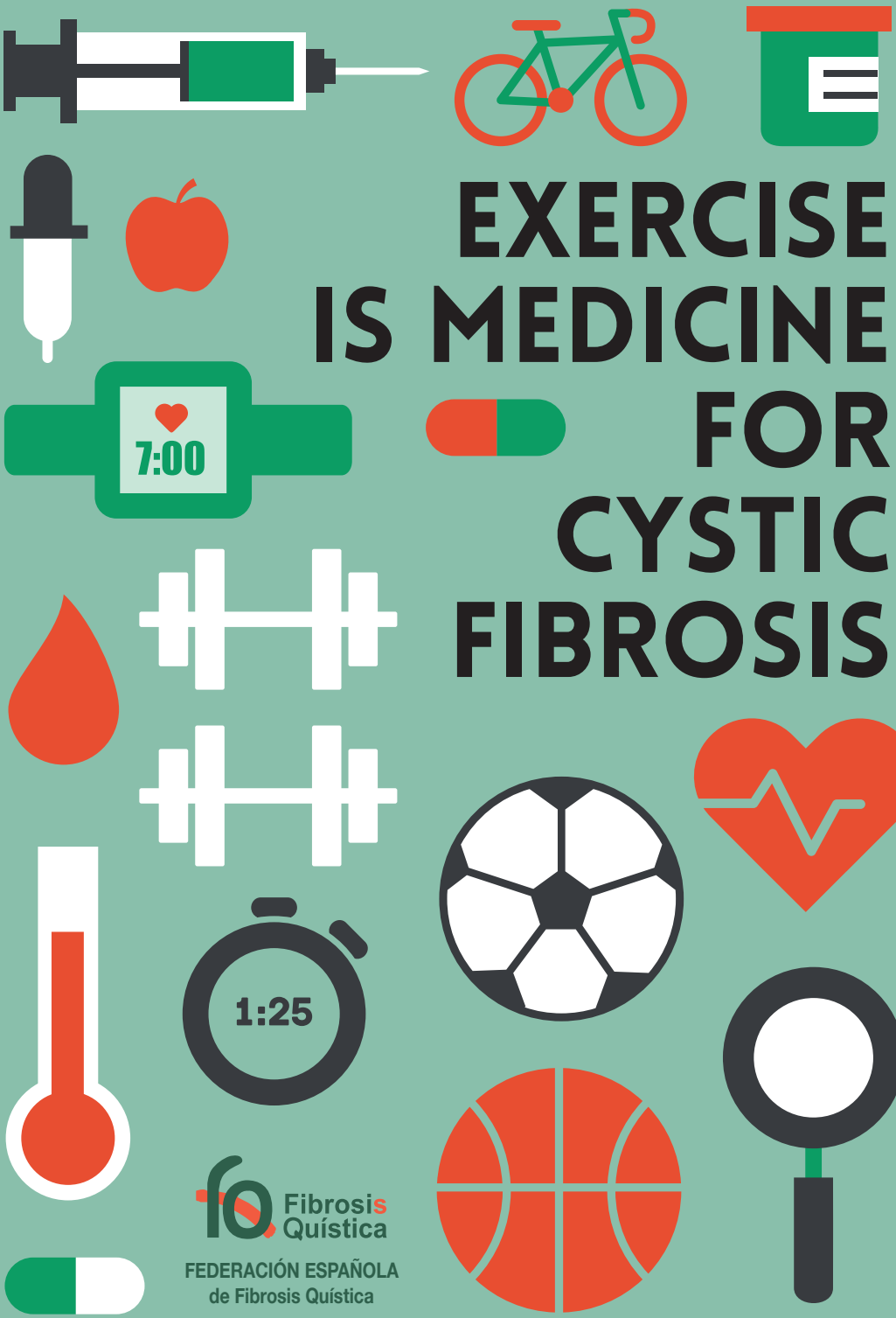
EXERCISE IS MEDICINE FOR CYSTIC FIBROSIS



Fibrosis Quística

FEDERACIÓN ESPAÑOLA
de Fibrosis Quística

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FEDERACIÓN ESPAÑOLA
de Fibrosis Quística

Doctoral School and Research
at the European University of Madrid

Faculty of Biomedical and Health Sciences

Faculty of Physical Activity and Sport Sciences
and Faculty of Biomedicine at the UEM

Faculty of Physical Activity and Sport Sciences
at the University of Castilla-La Mancha

Children's University Hospital "Niño Jesús" of Madrid

Coordinator and Editor Chief:
Margarita Pérez Ruiz, Ph.D.
Doctor in Medicine and specialist in Sports Medicine

Professor of Exercise Physiology
at the European University of Madrid

Collaborators:
José Ramón Villa, Ph.D.
Doctor in Medicine and specialist in Pediatric Neumology

Head of Neumology service at Children's University Hospital
"Niño Jesús" of Madrid (HUNJ)

Verónica Sanz Santiago, MPH.
Specialist in Pediatric Neumology

Dr. Luis Miguel López Mojares, Ph.D.
Doctor in Medicine and specialist in Sports Sciences

Dra. Susana Aznar Laín, Ph.D.
Doctor in Physical Activity and Sports Sciences

Dra. Elena Santana Sosa
Doctor in Physical Activity and Health Sciences

Translation: Ana Hill

PUBLISHER:
Spanish Federation of Cystic Fibrosis
C/ Duque de Gaeta, nº 56 - 14ª
46022 - Valencia
Tel.: 96 331 82 00
Fax: 96 331 82 08
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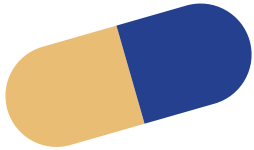

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What is essential is not to climb fast but to climb for a long time.

George Livanos

The important thing is not about what destiny holds for us but what can we do about it.

Florence Nightingale

PROLOGUE

In people with Cystic Fibrosis, exercise is able to achieve in the long term, adaptations and changes in such important organs as the lungs, heart, bones and muscle tissue, strengthening them and keeping them in a better condition. Therefore, it is already considered one of the basic pillars in the management of Cystic Fibrosis, along with the use of antibiotics, physiotherapy and nutrition. A sedentary behaviour is not a good attitude for anybody. Although if the rest of therapies are fulfilled, if exercise is not performed, we will not obtain the best benefits and the tissues will be in a worse condition.

The importance of including targeted physical activity and sport as a complement to the treatment of Cystic Fibrosis has begun to be understood just a few years ago, specially after publishing the study coordinated by Prof. Dr. Margarita Pérez, entitled "Prescription of an intrahospital physical exercise program, for children with Cystic Fibrosis. Effects on their functional capacity and general health", the study was awarded the "Pablo Motos grant 2009". For this reason, we believe that this guide is very innovative, since currently, there are no publications covering the aspects that we intend to treat here, providing specific guidelines about which physical exercises and sports that may be advisable to introduce a healthy lifestyle for people with CF (Cystic Fibrosis) and how to do them correctly.

The idea of publishing this guide has arisen directly from the suggestions made by young people and adults with CF to the Spanish Federation of Cystic Fibrosis, in order to have more information about physical exercise and the illness.

We have already done a short introduction about this subject on the DVD of aerosol therapy and physiotherapy in CF and now in this guide, we are going to develop it in greater detail. This will allow you to learn about the benefits of a basic and safe exercise to keep in optimal physical condition, something that will significantly improve the prognosis of the disease.

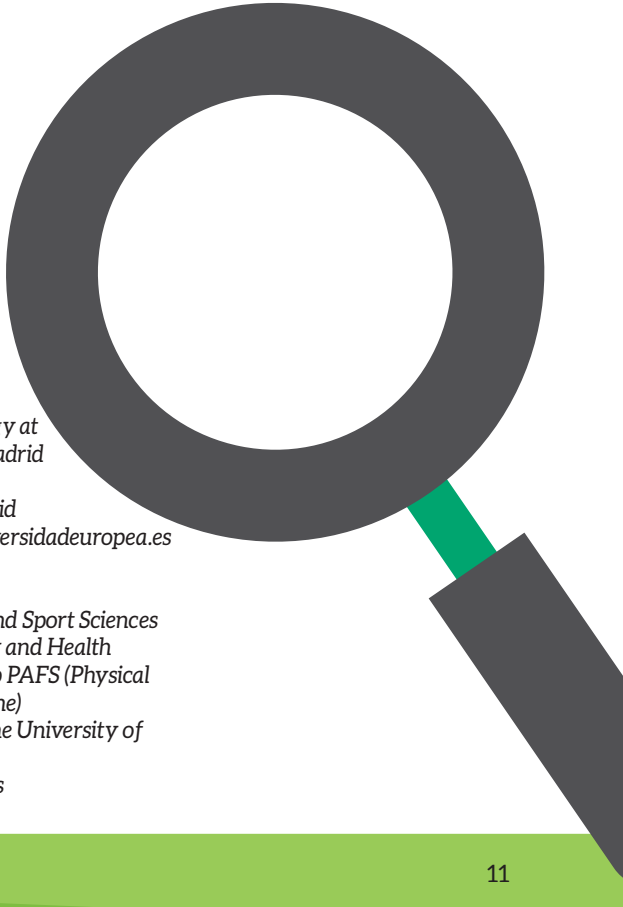
From the Federation, we want to contribute to this publication, in a conscious way, to establish a healthy lifestyle, specifically for people with Cystic Fibrosis and advancing to the ultimate goal of our Organization: improve the quality of life for people with CF and their families. All of this in addition to collecting and analyzing the most practical and concise information related to physical exercise for Cystic Fibrosis, in order to make some activities safer and useful for every day.

We aim to provide not only children and adults with CF, but also their families and professionals related to the disease, with a quality and reliable source of information, elaborated by specialists who, besides knowing in detail the topic, they completely understand the difficulties and problems associated with the illness. In these lines, we would like to take the opportunity to express our gratitude to the various authors for their collaboration and for the fantastic work they have done in writing this guide, which we are sure, will contribute to improving our lifestyle and making it healthier.

We hope you enjoy this interesting read, which should help to resolve any doubts and to understand the importance of including exercise in your daily routine, because health is the most important thing and if we overcome laziness, we will have moved one-step closer to overcoming the consequences of the disease.

Tomás Castillo Arenal.
President of the Spanish Cystic Fibrosis Federation.

00 INTRODUCTION



Margarita Pérez Ruiz
Doctor in Medicine and
specialist in Sports Medicine
Professor of Exercise Physiology at
the European University of Madrid
Doctoral School and Research
European University of Madrid
E-mail: margarita.perez@universidadeuropea.es

Susana Aznar Laín
Doctor in Physical Activity and Sport Sciences
Professor of Physical Activity and Health
Director of the research group PAFS (Physical
Activity for Health programme)
Faculty of Sport Sciences at the University of
Castilla-La Mancha (UCLM)
E-mail: susana.aznar@uclm.es

This guide attempts to explain the role of exercise in the treatment of Cystic Fibrosis. When we think of exercise, we should think about an activity that involves **some kind of stimulus**, since it is the basis for achieving improvements in the different organic systems and improvements that will allow us to develop the activities of everyday life with better quality. In addition, **this stimulus must be changed over time** to continue achieving improvements and adaptations that will allow us to feel better and better.

The expression ***"the exercise is good for your health"*** is a postulation so well known as a widely ignored and not only by the people with Cystic Fibrosis (CF) and their families but by the general population. That is why we currently have such high rates in all chronic diseases, that are in some way are adversely affected by the sedentary attitude of the population.

We have the opportunity to be able to count on a multidisciplinary team integrated by Sports Medicine Physicians, Medical Specialists in Paediatrics and Pneumology, University graduates and Doctors in Physical Activity and Sport Sciences. All of them are university professors who will try to convince us of the importance of exercise in our lives, if we want to have a good quality of life.

Once the basic concepts are known, the benefits and the minimum recommendations of exercise that you must do to stay in good physical condition, you need to incorporate this tool into your life. Therefore, that it will be easier to use most days.

Reasons for publishing this guide about exercise for CF

Human body has evolved to be physically active. In other words, our body needs exercise to stay healthy. Regular physical activity is associated with a better life quality. However, most adults, Spanish children and adolescents do not develop enough physical activity to achieve health benefits.

Physical inactivity during the first years of life is presently recognised as an important contributory factor in increasing the levels of obesity and other serious medical disorders seen in children and adolescents in Europe and other parts of the world. In particular, for Cystic Fibrosis (CF) it has been observed despite the lung function decreasing due to the pathology, physical exercise can keep the functional capacity and as a result maintain or improve muscle mass, that contributes positively to the maintenance and/or improvement of physical fitness and quality of life of the patient.

Spontaneous physical activity is scarce and sedentary leisure is increasing. It is therefore essential to make an effort to "reintroduce" physical activity in our lives. This guide aims to contribute to this task.

The improvement of the levels of physical activity in children and adults with CF is the responsibility of all members of society. However, as a person who interfaces directly with this pathology, you constitute a particularly important component of the network of influences. Schools, health centres (GPs), hospitals, households and communities are the places that offer assistance to children and adults with CF to improve their physical activity.

Let's help the people with Cystic Fibrosis to be more active!

Who this guide is aimed at

This guide has been designed to provide information to adults who work with children, teens and adults with CF, on the importance of exercise for this sector of the population and how you can help people with Cystic Fibrosis to be more active. Also, to show how to promote in an efficient way, the physical activity in the presence of this disease. This is a document relatively detailed that has been developed to provide with useful information to adults with advanced

knowledge of physical activity (teachers, educators, coaches, health care professionals, etc.) and also to those who have limited knowledge of the subject.

As far as possible, we have tried to avoid presenting an overly complicated text. However, at the end of this guide you can find a list of more extensive and scientific sources of information if you are interested.

Objectives of this guide

The Guide has been designed on the basis of the following principal objectives:

1. Improve the understanding of the concepts and relevant issues in relation to physical activity and fitness or physical condition.

2. Provide information on the importance of the physical activity for people with CF, during the years of childhood, adolescence and adult life, explaining the benefits that the human body gets by doing physical exercise.

3. Describe the current recommendations on physical activity in children and young people and how to perform them.

4. Clarify how to develop a proper physical exercise program, whether we are in a sports centre with a lot of material or if we do not have any material.

Learn the tools that lead you to know how to carry out exercise, if you get the skill, you will generate adherence to exercise.

Go step by step, do not rush, so your body will make the necessary adjustments that will let you go another step further,

Go step by step.
You might take longer, but you will probably have more success.

until you find yourself comfortable and you will realise that exercise is now part of your life. Then you will have achieved the goal and you will have received your reward.

Changes of behaviour, only you are responsible for them, no one else can do that for you and as we said before "Go step by step. You might take longer, but you will probably have more success".

The physical exercise is a very useful and safe tool for CF. In the long term, physical exercise can achieve several adaptations and changes in organs such as the lung, heart, muscles and bone tissue. Strengthening these tissues is very important, keeping them as healthy as possible is your responsibility while waiting for a genetic solution, which is still pending. If you do not do exercise, even if you fulfil the rest of therapies, you will not get the best results and all your tissues will be in worse condition. Sedentary behaviour is not the best attitude for anyone.

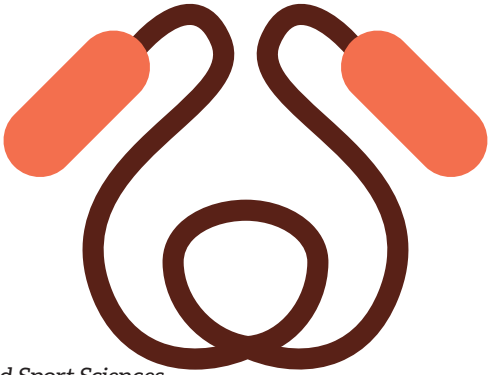
It is our responsibility to use the stimulus of exercise to activate mechanisms that strengthen our organic systems and thus wait in the best conditions for those drugs that enhance and correct the chloride channel function (CFTR).

While waiting for new drugs, keep yourself physically active.

The main objective of this guide is to contribute to the improvement of the levels of physical activity in people with Cystic Fibrosis.

01

GENERAL CONCEPTS OF PHYSICAL ACTIVITY AND FITNESS



Susana Aznar Laín
Doctor in Physical Activity and Sport Sciences
Professor of Physical Activity and Health
Director of the research group PAFS
Faculty of Sport Sciences at the University of Castilla-La Mancha (UCLM)
E-mail: susana.aznar@uclm.es

Concepts and definitions

Physical Activity: is defined as a body motion produced by voluntary muscular action, which increases the energy expenditure. It is a broad term that encompasses the concept of "physical exercise".

Physical exercise is a more specific term that involves physical activity planned, structured and repetitive performed with a goal, which is improving or maintaining the physical condition of the person. For example, the activities of gardening or climbing stairs at home cannot be classified as structured 'exercise', but clearly, they are physical activities.

Physical condition or "fitness" refers to a physiological state of well-being that provides the foundation for the tasks of everyday life, a level of protection against chronic diseases and the basis for the development of sports activities. Essentially, the term physical condition describes a set of attributes related to the performance of the person in the area of physical activity.

Relevant descriptors of activity and physical exercise

The "dose" of exercise that a person receives depends on factors included in the principle FITT (frequency, intensity, time, and type):

- Frequency (level of repetition): the number of times that the person carries out physical activities (often expressed in number of times a week).
- Intensity (level of effort): the level of effort that involves physical activity (often described as light, moderate or vigorous).
- Time (duration): the duration of the session of physical activity.
- Type: the specific category of exercise that person performs (for example, running, swimming, etc.).

These factors can be manipulated in order to vary the "dose" of exercise. Frequently, this dose is expressed in terms of energy expenditure (calories burned). It can be seen that, if physical activity is more intense, the person burns calories at a higher rate, which can reduce the time required to burn a set amount of calories.

Physical exercise recommendations

Current practice recommendations are as follows:

Recommendations on physical activity for children and adolescents:

1. Children and adolescents must do 60 minutes (and up to several hours) of physical activity at moderate to vigorous intensity, all or most of the days of the week.
2. At least two days a week, this activity should include exercises to improve bone health, muscular strength and flexibility.

Recommendations on physical activity for adults:

1. Adults should accumulate a minimum of 150 min (2 hours and 30 minutes) of moderate physical activity per week.
2. If not, 75 min (1 hour and 15 minutes) of vigorous physical activity per week.
3. A combination of both 1 and 2, would be ideal.

Clarifications concerning moderate physical activity

Among all of the factors included in FITT, the intensity is probably the most difficult to measure. General recommendations on physical activity for children, adolescents and adults, refer to the importance of doing at least moderate intensity exercise.

People who usually carry out moderate-intensity activity might feel:

· An increase in breathing rate that does not hinder the ability to speak.

· An increase in heart rate to the point that it can be easily felt in the wrist, neck, or chest.

· A sensation of increased temperature, possibly accompanied by sweating on hot or humid days.

A session of moderate intensity activity can be maintained for many minutes and does not cause fatigue or extreme exhaustion in healthy people, when it takes place over a long period.

It is important to understand that the moderate intensity has a relative character with respect to the level of physical condition of each person. For example, a person with a better fitness level must perform an intensive activity with higher intensity than someone with worse physical condition, in order to feel similar sensations of increased breathing rate, heart rate and temperature, which are the characteristics of physical activity of moderate intensity.

Below we describe in detail various methods to evaluate the exercise intensity.

Additional methods to measure the intensity of the physical activity

There are plenty of ways to evaluate the intensity of physical activity. The most used methods are outlined as follows:

1. The trial of the "talk test"

The test of the ability to speak, in order to measure the exercise intensity, is simple:

· Light intensity: a person who performs a light intensity physical activity must be able to sing or have a conversation while carrying out the activity. Some examples of light intensity activity are walking and cleaning.

· Moderate intensity: a person who performs a physical activity of moderate intensity should be able to hold a conversation, but with some difficulty, while carrying out the activity. Examples of moderate-intensity physical activity may be brisk walking, cycling or dancing.

· Vigorous intensity: If a person gasps or gets out of breath and cannot maintain a conversation easily, the activity can be considered as vigorous. Examples of vigorous activity are jogging or sports, such as basketball, handball, swimming, etc.

2. Heart rate

Heart rate can be measured on the wrist (radial pulse) or in the neck (carotid pulse) and must be converted into the number of beats per minute of the heart ("beats per minute" - bpm). If you don't have a heart rate monitor, you can measure your heart rate during a full minute or you can measure during a shorter period of time (for example, 15, 20 or 30 seconds) and multiply the value obtained by the relevant factor (4, 3 or 2, respectively) in order to convert it into beats per minute.

Knowledge of resting heart rate and maximum heart rate of the person is required to measure the intensity of the workout more efficiently. The heart rate at rest is best measured when the person is fully rested, for example, when wakening up in the morning or after sitting quietly for several minutes. The maximum heart rate, if we do not have the actual data measured on an exercise stress test, is calculated by using the simple equation of "220 - age". For example, if a person is 15 years old, your maximum heart rate estimated it would be 220 - 15 = 205 bpm.

The best method to determine the ideal heart rate when assessing the intensity of physical activity consist in the use

of the technique known as "Method of Heart Rate Reserve" (HRR), also known as the "Karvonen method".

In this method, the resting heart rate (HRrest) is subtracted from the maximum heart rate (HRmax), in order to get the heart rate reserve (HRR). For example, the person of 15 years mentioned above, with a heart rate at rest of 80 beats per minutes (bpm). Heart rate reserve (HRR) of this person is as follows: HRmax (205) - HRrest (80) = 125 bpm.

In order to calculate a range of heart rates for practical purposes, consult first table 1 listed below to determine the percentage values (%) of the heart rate reserve (HRR).

We can observe that the moderate intensity is corresponding of values of 40% - 59% heart rate reserve = 50 (0.40 x 125) - 74 (0.59 x 125). We must now add the resting heart rate to each number in order to determine the Target Heart Rate. Therefore, the corresponding Target Heart Rate or Training Heart Rate Zone (THR) for young person cited in our example is 130 (50+80) to 154 (74+80) bpm.

For the exercise of vigorous intensity, for this young man, THR would be 155 to 185 using the methods described before.

Type of Intensity	Relative intensity	
	% Of the heart rate reserve (% HRR)	Rating of perceived exertion (scale of perceived effort - RPE)†
Very light	< 20	< 10
Light	20-39	10-11
Moderate	40-59	12-13
Strong (vigorous)	60-84	14-16
Very strong (very vigorous)	>85	17-19

Adapted from Med Sci Sports Exerc 1998, 30:975-991.
† see section 3, appearing later, where it clarifies the concept of valuation of the perceived exertion.

Table 1: classification of the intensity of physical activity using the heart rate reserve percentage and the rating of perceived exertion.

3. The evaluation of the perceived exertion using the Borg perceived exertion scale

The perceived exertion is based on the physical sensations a person experiences during physical activity. An example of the scale of Borg is shown in Figure 1.

During exercising, it should be observed the expressions of the scale of assessment, evaluating the subjective perception of effort as sincerely as possible, and thus obtaining the corresponding figure. This is the rating of perceived exertion or RPE (rating of perceived effort).

As shown in Figure 1 outlined above, a moderate-intensity physical activity is represented as RPE value between 12 and 13 on the Borg Scale (around the description "Somewhat Hard"). Light intensity to vigorous activities, are represented between 10-11 and 14-16, respectively. Your heart must beat

6	No effort/No Exertion At All
7	Extremely Light
8	
9	Very Light
10	
11	Light
12	
13	Somewhat Hard
14	
15	Hard (Heavy)
16	
17	Very Hard
18	
19	Extremely Hard
20	Maximal Exertion/Maximum Effort Possible

Figure 1: Borg Rating of Perceived Exertion Scale

at 120-130 bpm and your perception of effort should be 12-13 (you must familiarize yourself with the scale and learn to recognize such perception).

4. MET Level (metabolic equivalent - MET - level)

A MET is defined as oxygen uptake in ml/kg/min.

A metabolic equivalent (MET 1) is the amount of energy (oxygen) used by the body at rest while sitting quietly and calm. The intensity can be described as a multiple of this value. The harder your body works during the physical activity, the more oxygen is consumed and the higher the MET level.

· Any activity that burns 3-6 METs, is considered moderate-intensity physical activity.

· Any activity that burns > 6 METs, is considered vigorous-intensity physical activity.

Typed tables that define the physical activity and MET levels can be used in order to assess approximately the intensity of such activity, as for example, Table 2 that appears below:

Activity	*Intensity Level*	*METs Level ml/kg/min*	*Energy expenditure expressed in Kcal* *(for a person who weighs 30 kg, doing the activity during 30 min.)*
Ironing	*Low*	*2.3*	*35*
Cleaning and dusting	*Low*	*2.5*	*37*
Walking 3 - 4 km/h	*Low*	*2.5*	*37*
Painting	*Moderada*	*3.0*	*45*
Walking 4 - 6 km/h	*Moderada*	*3.3*	*50*
Vacuuming	*Moderada*	*3.5*	*53*
Playing golf (walking, carrying clubs)	*Moderate*	*4.3*	*65*
Badminton (for fun)	*Moderate*	*4.5*	*68*
Playing Tennis (doubles)	*Moderate*	*5.0*	*75*
Walking >6 km/h	*Moderate*	*5.0*	*75*
Mowing the lawn	*Moderate*	*5.5*	*83*
Cycling at 16-19km/h	*Moderate*	*6.0*	*90*
Dancing intense	*Intense*	*6.5*	*93*
Cycling at 19-22km/h	*Intense*	*8.0*	*120*
Swimming (front crawl 45 meter/min)	*Intense*	*8.0*	*120*
Playing tennis (individual)	*Intense*	*8.0*	*120*
Running at 9-10km/h	*Intense*	*10.0*	*150*
Running at 10-12km/h	*Intense*	*11.5*	*173*
Running at 12-14km/h	*Intense*	*13.5*	*203*

Source: Ainsworth et al. [4]

Various components of the physical activity

Of course, there are many different types of physical activity used to develop various aspects of the physical condition. The most important types of physical activity for children and young people's health are:

1. Activities that involves the cardiovascular work (aerobic).
2. Activities related to the strength and/or the muscular resistance.
3. Activities related to flexibility.
4. Activities related to coordination.

1. Activities that involve cardiovascular work (aerobic)

Cardiovascular activities are called with frequency "cardio respiratory" or "aerobic" activities because they require the body to transport oxygen using the heart and lungs. Cardiovascular endurance is the ability of our body to carry out tasks that require the use of large muscle groups, generally for periods of time relatively long (several minutes or more). To perform an exercise of resistance, our heart and our lungs adapt in order to be more efficient and supply the working muscles with oxygenated blood that they need to perform the task.

You can improve the cardiovascular endurance through the performing of ongoing activities, such as walking, running, swimming, cycling, paddling a canoe, dancing, etc.

When performing such activities it is important to remember:

· That must progress in a reasonable way: if you have not done before any kind of activity, you should start exercising gradually with intensity and duration relatively low and increase them gradually as your physical condition improves. Remember that the intensity is relative and must be well monitored (see section intensity).

· The selected activity should be fun and accessible: this fact will increase the odds of carrying it out regularly. If you can not enjoy the activity, if it requires a lot of expensive equipment, or if it requires long journey to do it, it will be less likely that the person will continue with the activity.

· Security issues: include aspects such as the use of appropriate safety equipment (for example, helmet when cycling) and the prevention of injury, for example: wear the proper shoes, drink water before, during and after activity, use of sunscreens if we are exposed to the sun, etc.

2. Activities related to the strength and/or the muscular resistance

Muscle strength is the ability of the muscle to generate tension and overcome a counterforce.

Muscular resistance is the ability of the muscle to maintain its tension or the contractions during a prolonged period of time. These activities serve to develop and strengthen the muscles and bones. We use the strength and muscular endurance when we push, pull, raise, transport or carry things such as heavy shopping bags.

The activities of strength and muscular endurance/resistance can be done:

· With the weight of the person (jumping rope, climbing, press-ups, etc).

· With the weight of a companion (wheelbarrow walk, tug of war, wrestling with a friend, etc).

· Or with activities such as throwing a ball, paddling a canoe, rowing, weightlifting in a gym, carrying objects, etc.

When carry out strength and muscular endurance/resistance activities, the following criteria must be taken into account:

· It must be carried out in a reasonable way: If you are a beginner to this type of activity, start slowly, learning the technique and posture with a low resistance, in order to avoid aches and muscle injuries.

· For the strength activities is not necessary always to use weights: there are many activities that test the muscle strength without use them. Examples include activities where you support your own body weight, such as arms, climbing, gymnastic activities, etc. Other simple workouts also used to develop muscular strength are the changes of planes of motion (range-of-motion exercise), the use of resistance bands, the strength of a teammate, rubber-tubing exercises, etc.

· If you have doubts, it is always a good idea to ask an expert, such as a teacher of physical education, a specialized fitness trainer, a doctor of sports medicine, etc.

3. Activities related to flexibility

Flexibility is the ability of the joints to move in their full range of motion. Flexibility has specific characteristics for particular parts of the body and depends on the type of joint or joints implicated, also the elasticity of the muscles and the connective tissue (e.g., tendons and ligaments) that surround the articulation or joints. Flexibility is beneficial for all activities related to push-ups, movements, contortions, extensions and stretches.

Some activities that improve flexibility are gentle muscle stretching, sports like the gymnastics, karate, martial arts, mind-body activities like yoga and Pilates and any activity of strength or muscle endurance when the muscles are working in their full range of motion.

When doing activities of flexibility is important to remember that:

· You must be patient. It takes time to achieve improvements in terms of flexibility, often several weeks or even months.

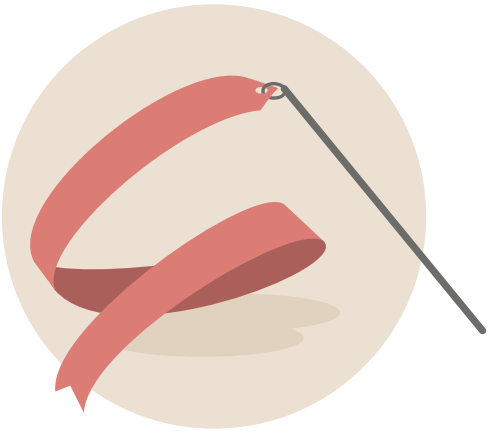
· You should never stretch to the point where it becomes painful and movements always must be carried out with control, no bouncing or pulling. NEVER force yourself to imitate another person who is more flexible than you are. The only thing that you will achieve is an injury!

· Regular stretching should be performed (preferably several times a week or even every day). The reasons justifying this periodicity are that you lose flexibility level easily if you do not stretch regularly to maintain it, good flexibility can help to prevent injuries and that the flexibility decreases as our age increases.

· It is good idea to start practicing flexibility exercises at an early age (when we are more flexible) and continue doing them all your life.

· It is better to stretch your muscles and joints when they are warm, because they are more flexible. Therefore, the best time for stretching could be after a physical activity as part of cooling down.

In addition, it is important to know that the flexibility is different for girls and boys (often better in the case of girls) and during the main stages of growth, it is usual to observe significant reductions of flexibility, due to bones grow faster than muscles and tendons.



4. Activities related to coordination

Motor coordination is the ability to use the brain and the nervous system along with the motor system in order to carry out smooth and precise movements. Coordination activities include:

· Body balance activities such as walking on a balance beam or standing on one leg.

· Rhythmic activities such as dance.

· Activities related to kinesthetic awareness and spatial coordination, such as learn how to perform a somersault or a new dance step.

· Activities related to coordination between eye and foot, such as kicking a ball or dribbling in football.

· Activities related to the coordination between hand and eye (eye-hand), such as racket sports, or throwing a ball and catching it.

You should take into account the following criteria in relation to coordination activities:

· Be careful to avoid falls and other accidents, because children are often so absorbed by these activities, that they lose awareness of their surroundings.

· Coordination activities are excellent for motor development, especially for children. Most children love them!

· The learning curve of these capabilities is different for each child: some learn faster than others do.

Key points

· Physical activity is a bodily movement produced by muscle-skeletal traction that requires energy expenditure.

· Dosage (or amount) of physical activity is a combination of frequency, intensity, time and type of activity (FITT).

· For a continuous improvement of physical condition, a gradual and progressive increase of physical activity should be applied.

· The intensity of physical activity can be measured through the "Talk Test", the Heart Rate, the Scale of Perceived Effort and the Metabolic Equivalent (MET).

· The most important types of physical activity to achieve a health-related fitness are cardiovascular activities, muscular strength/endurance (or resistance), flexibility and coordination.

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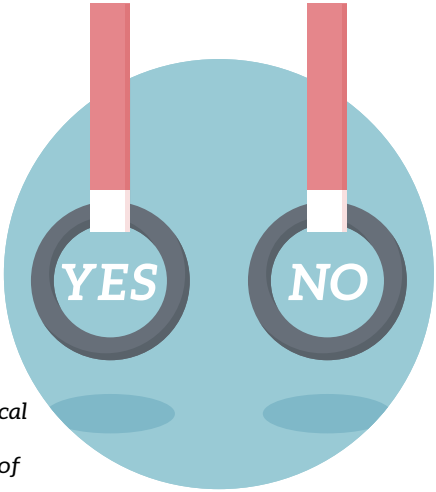
02

DO CHILDREN WITH CYSTIC FIBROSIS FOLLOW THE RECOMMENDATIONS FOR PHYSICAL ACTIVITY?



Susana Aznar Laín
Doctor in Sports Medicine and Physical Activity
Professor of Physical Activity and Health
Director of the research group PAFS (Physical Activity for Health programme)
Faculty of Sport Sciences at the University of Castilla-La Mancha (UCLM)
E-mail: susana.aznar@uclm.es

The current recommendations for physical activity to maintain or improve health (see chapter 1), suggest that children should accumulate a minimum of 60 minutes of MVPA a day. However, many healthy Spanish children do not achieve this minimum level of physical activity.

This situation is even more worrying for children with Cystic Fibrosis. Regular physical activity and aerobic training (see Chapter 1 Cardiovascular Activities) are very important in the design of the program of physical exercise for people with CF, because they help to reduce the loss of lung function and improve the quality of life. Doing regular physical activity and aerobic training are very important because they are predictors of survival, i.e., if the person with CF is trained (determined with a stress test peak oxygen consumption), they will live longer and have a better quality of life.

That is why it is very important to quantify how much movement people with CF do, in order to check if they achieve the minimum recommendations of physical activity.

How can I measure the level of physical activity?

Physical activity levels, in children and adolescents, can be measured using a number of different methods, each of them with its own advantages and limitations. Several objective techniques involving the measurement of the heart rate can be done (using a "heart rate monitor"), the accelerometry (using small sensors of movement usually attached to the hip or wrist) and pedometers (devices that measure the number of steps). These techniques can be used in a large number of subjects, but all of them have the disadvantage of requiring a significant level of compliance by subjects (wearing it, which is a small inconvenience).

Finally, several subjective techniques can be used, such as self-reports, questionnaires, interviews and diaries. These techniques have the advantage of being quite affordable and easy to use with large samples, but this might experience

the problems inherent to memory errors and the subjective nature of the auto-report. Children, in particular, have difficulty remembering exactly what their activities were in the days or the weeks prior.

Currently, the most appropriate method in research to measure physical activity is the accelerometer. However, it is an expensive and research-oriented instrument. A pedometer is very useful for everyday use. With it we can review the steps we take every day, have feedback of our movements and compare them. For example, using the pedometer three consecutive days and estimating the average steps per day. From this data it is proposed a weekly increase and evaluation. There also exist references for adults and children about how many steps per day are recommended for health benefits: a minimum of 10,000 steps per day for adults and values around 12,000 steps a day for children.

Why are children less active today?

It has been estimated that today, children spend approximately 600 kcal a day less than 50 years ago. There are multiple factors for this including the following:

1. More sedentary leisure activities, such as watching television, Internet and computer games, instead of playing outside.

2. Less physical education at school.

3. Fewer opportunities to develop leisure activities.

4. The increase in motorised transport (for example, the cars), in particular to go to school.

5. The increase in urbanisation, which does not promote the active and safe transportation such as walking or cycling.

6. The increase of mechanization in society (lifts, escalators, etc.).

7. An environment (home, school, society) that does not promote physical activity (for example, obese and inactive parents).

Do the children with Cystic Fibrosis follow the minimum recommendations for Physical Activity?

There is a scientific study where measured levels of PA with accelerometers in Spanish children with CF. In this study they compared the levels of physical activity and aerobic fitness between two groups of children of the same age and gender, one group of patients with CF and another healthy control group. Of 39 CF patients enrolled in this study, only 2.1% of them met the recommendations of PA, while 34.2% of the control group of healthy children did. As we can see, half (50%) of the healthy control group met the minimum recommendations of PA, but children with CF were surprised by his almost nonexistent compliance. Curiously when analysing the data, children with CF had more quantity of light physical activity than healthy children, but much less physical activity corresponding to moderate and vigorous. In addition, the aerobic fitness, also measured in this study, was correlated with intensive physical activity or moderate to vigorous intensity.

Key Points:

· Therefore, it seems very important to promote children moving more.

· That the PA of moderate to vigorous intensity is better rather than light intensity physical activity.

· That physical activity moderate to vigorous is very important, because it is the activity that improves aerobic fitness.

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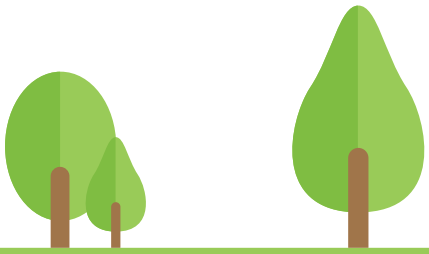
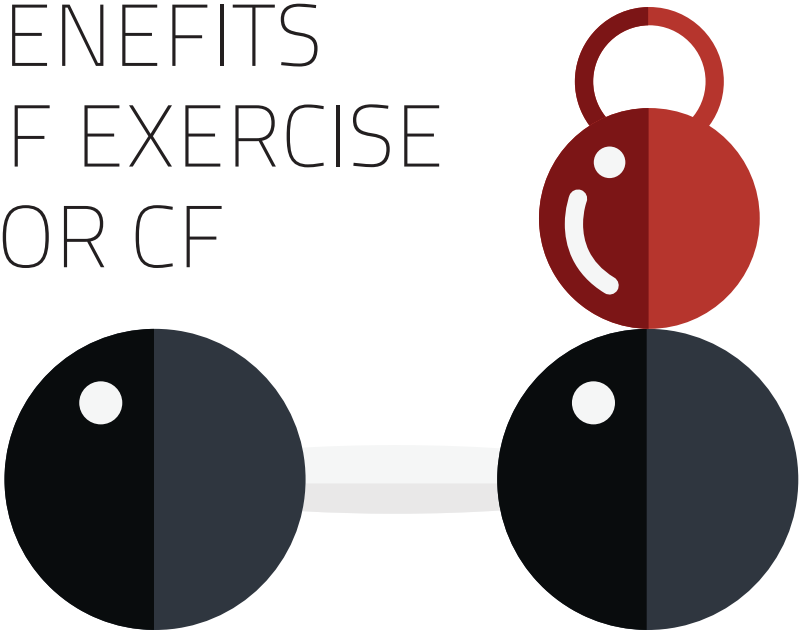


03

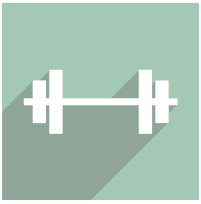
SPECIFIC BENEFITS OF EXERCISE FOR CF



Margarita Pérez Ruiz
Doctor in Medicine and specialist in Sports Medicine
Professor of Exercise Physiology at the European University of Madrid
Doctoral School and Research
European University of Madrid
E-mail: margarita.perez@universidadeuropea.es

Specific benefits of exercise for CF

Intolerance to exercise is an established feature of CF and is dependent on the type of mutation, on the state of muscle mass conservation, on the pulmonary system and on the maintenance of the physical condition. Exercise is one of the tools to keep the muscular mass in better shape, the pulmonary system and physical condition.



Why use exercise in CF?

Because there is scientific evidence for more than four decades that inform us about the great benefits provided by this tool for CF.

Exercise still in this century, is a tool very little used but highly recommended by the official agencies responsible for ensuring the health of the population.

Canadian Paediatric Society / Société canadienne de pédiatrie

Recommends **exercise in children with chronic diseases** such as: juvenile idiopathic arthritis, haemophilia, asthma and CF.

The majority of researchers attribute the benefits of exercise in these patients, to the improvement in cleaning the mucus from the respiratory tract; increased strength and the resistance of ventilatory muscles; reduction in airway resistance; increase in tolerance to exercise and increase the feeling of well being.

At the end of the 70´s and the beginning of the 80's, Keens et al. (1977) and Zach et al. (1981) showed that physical training programs can be used to improve ventilation and help to clear the mucus from the respiratory tract.

The Group Kruhlak et al. (1986) also found that there was a reduction in trapped air after exercise, especially in the apical regions of the lungs. Other researchers have found reductions in the residual volume after general training programmes *(Andreasson B, 1987 O'Neil PA, 1987)* or specific strength programs *(Strauss GD, 1987)*.

In addition, the increase of physical activity is accompanied by a slowdown in the deterioration of lung function *(Schneiderman-Walker J, 2000)*.

The peak oxygen uptake (VO_2max) and peak power output (Wpeak) (PPO) and the tolerance to exercise have experienced drastic improvements in the various studies carried out in patients with CF especially among patients with a degree of major disorder.

A better aerobic power (VO_2max) is associated with fewer hospital admissions for pulmonary exacerbations (Pérez M. et al. 2014)..

The progressive nature of the disease and the need of the patients to be under permanent treatment, make people with CF and their closest relatives suffer from a situation of continuous anxiety, on which physical activity can be effective.

Children with moderate-severe CF can benefit from aerobic and strength training.

In the long term, despite the drop in the pulmonary function, it has been observed that people with CF moderate pulmonary dysfunction, can with exercise keep functional ability

measured through the VO_2 Peak, as a consequence of maintaining an improved muscle mass, nutrition and growth, contributing in a positive way to the maintenance of fitness (Stranghelle JK, Skyberg et al.)

> **The good news** is that the exercise carried out continuously manages to improve the physical condition of the person with CF.

The short and long term benefits of exercise on fitness are reflected in the improvement of the quality of life, in the psychological development and in the increase of aerobic power, which is a variable of the disease prognosis.

> Good physical condition is associated with a better prognosis of the disease.

> **Exercise is a low cost medicine.**

General benefits of exercise for CF

Genetically, people of the 21st century continue being citizens of the Palaeolithic, so those who engage in a more active lifestyle will live better and longer, suffering from reduced risk of chronic diseases.

The Spanish Association of Paediatrics alerted that 1 of every 4 Spanish adolescents do not perform any physical activity and just 10% meet the recommendations of doing at least one hour of exercise a day.

The risk induced by physical inactivity is similar to those produced by high blood pressure, high cholesterol and obesity and according to some authors, it is comparable to the risks linked with smoking.

It is required a minimum dose of exercise to improve health. The benefits are greater when more time is devoted to maintaining the effective dose of exercise. Regular and continued physical activity can lead to a significant reduction of the mortality rate.

Goraya and his group (2000), in a follow-up of more than 3,000 subjects over 6 years, suggested that an increase of 1 MET would result in reductions in the cardiovascular risk of 14% among the youth population and 18% among those older than 65 years. Similar results have been obtained by other groups *(Myers, 2002;) (Gulati, 2003)*.

Analysing large populations (more than 12,000 males and more than 2,300 women) undergoing cardiopulmonary rehabilitation, when increasing 1 $mL \bullet kg^{-1} \bullet min^{-1}$, in the peak of oxygen (VO_2 Peak), this leads to a reduction in mortality from cardiovascular causes of 10%. It has also shown that for each 1% increase in VO_2 Peak from training, obtains a reduction of 2% of cardiovascular mortality. Stay more than two hours sitting increases the risk of cardiovascular disease *(T.D Katzmarzyk et al. 2009)*.

Physical exercise can delay the onset of diabetes by increasing the manifestation of determined genes involved in the regulation of glucose.

Exercise improves the performance of the GLUT-4 receptors that help introduce glucose inside the cells, keeping blood glucose levels controlled.

Diabetic patients also benefit from exercise and the amount of insulin administered should be amended depending on the performed exercise.

In subjects with type 1 diabetes, the exogenous contribution of insulin will allow the control of the disease. The aim is to avoid as far as possible the hyperglycemias, since, in the long term; they produce an alteration of blood vessels in many parts of the body accumulating glucose in some of its components. This fact has fatal consequences for the organs affected such as the kidney, eyes, nerves, etc. Insulin should be administered in a way that mimics our pancreas the best way possible: release insulin into the blood after each ingestion and limiting their release both in periods of fasting or during exercise. Therefore, insulin should be administered according to the dietary habits of the subject.

The muscle cells during exercise capture blood glucose, without the necessity for insulin. This contributes to decreased blood glucose even in the absence of insulin. Therefore, exercise is one of the pillars of the treatment for diabetes with insulin and diet. It is necessary that the diabetic patient knows perfectly these mechanisms when it comes to exercise, since if they are not taken into account, may appear a hypoglycaemia as a result of combining both factors: administration of exogenous insulin and exercise.

Why isn't there risk of hypoglycemia in a healthy person when exercising? Simply because the pancreas limits release of insulin during exercise.

Chronic inflammation is in the pathogenesis of pathologies such as insulin resistance, atherosclerosis, neurodegeneration, and tumour growth. Continued exercise protects against type 2 diabetes, cardiovascular disease, colon cancer, breast cancer and dementia.

The works of Petersen AM, 2005 suggest to us, that as inactivity and obesity generate an increase of proinflammatory cytokines released to circulation from adipose tissue and that condition produce a cascade of cytokines in blood that originates a systemic state of low intensity inflammation which is related to the onset of atherosclerosis, insulin resistance, neurodegeneration and some cancers. When doing regular physical exercise, muscle contraction causes the appearance of cytokines inhibitors of that state of low intensity inflammation; we could call them anti-inflammatory cytokines. In addition, there are many other cytokines generated with beneficial effects on the use and oxidation of lipids, glucose consumption, or the distribution of visceral fat, achieving as well the increase of substances that regulates growth and neuronal survival.

Exercise contributes to neuronal regeneration by stimulating the production of a neurotrophic factor and maintains better brain irrigation, improving emotional and mental performance.

The power of our cells to repair themselves, depends on the length of their Telomeres; their length reduces as cell undergoes cell divisions, each cell has a clock that tells how log it has to live. Depending on our lifestyle, we keep our Telomeres most preserved. Regular physical exercise delays the shortening of Telomeres by activating the cellular defence for more time. Accumulating errors in behaviour from an early age (nutritional or deficit patterns of movement) increases cholesterol, increase of glucose, among other substances, causing endothelial damage which is reversible, since the cell is able to repair the damage by activating its cell divisions that repairs the damage. If this process is activated from an early age, cell will soon spend the length of their Telomeres that enables them to defend against damage, arriving a time and an age when the process is not reversible and cells are no

longer able to defend and repair themselves, opting to release tissue factors that obstruct the artery, this process affecting the smaller vessels of the brain.

They are regions of non-coded DNA, whose main function is the structural stability of chromosomes in eukaryotic cells, the cell division and the lifetime of the cell lines.

There is no drug that provides global health benefits like physical exercise... "Everything that gets worse with age, improve with exercise". Along these lines, we have to consider physical activity and exercise as health tools, not only to prevent but to cure and which therefore should be included in the medical paradigm for the 21st century.

The Spaniards spent more than 9 hours a day sitting down, very recent scientific studies warn about the negative effect of this conduct.

Platon already said: lack of activity destroys the good condition of every human being, while movement and methodical physical exercise preserve and retain it.

Sedentary lifestyle:
Too much risk, few benefits.

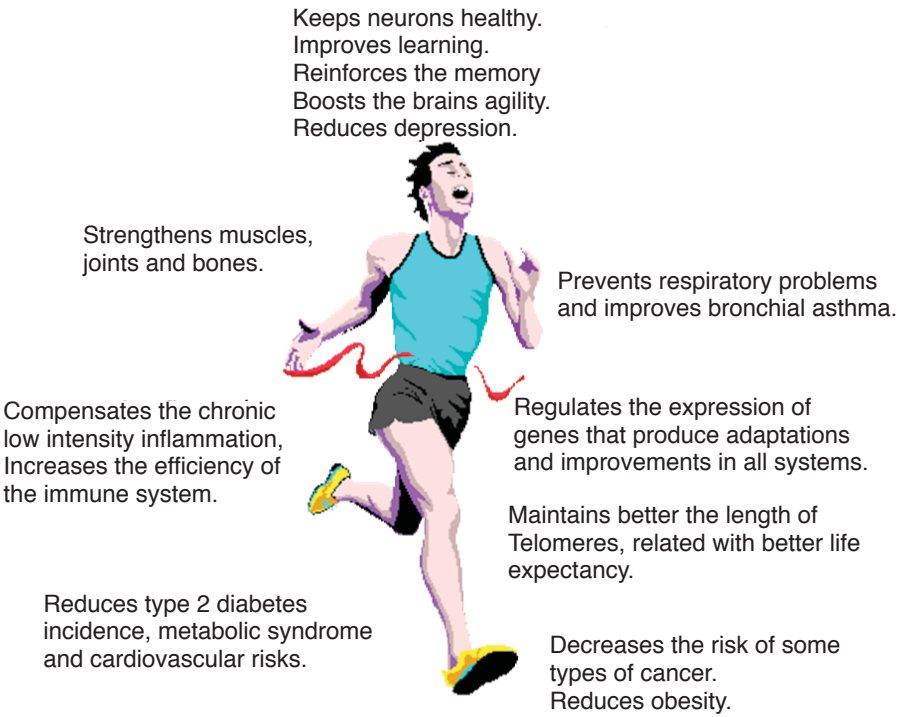


Fig. 1 Resume of exercise benefits for health.

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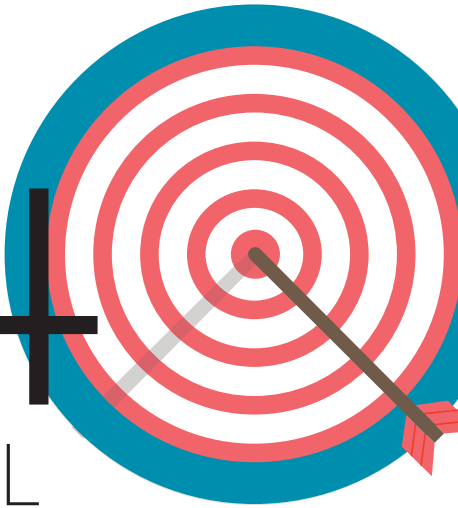
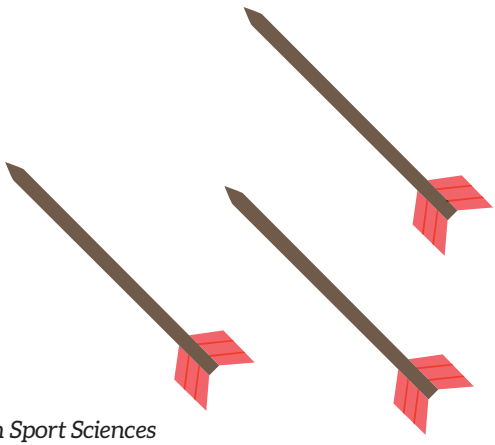


FUNCTIONAL ASSESSMENT OF CHILDREN WITH CYSTIC FIBROSIS



Luis Miguel López Mojares
Doctor in Medicine and specialist in Sport Sciences
Faculty of Biomedical Sciences and Health
European University of Madrid
E-mail: lmiguellopez@universidadeuropea.es

Aerobic power (VO_2 Peak) and anaerobic capacity (Anaerobic threshold)

For people with Cystic Fibrosis it is especially important the proper evaluation of the organism response to a given effort, in order to do physical exercise safely.

General training principles are applicable to any person, while patients in general and people with Cystic Fibrosis in particular, require greater precision in order to prescribe the dosage of exercise suitable for each person, allowing the best benefits and minimizing risks. For this reason, the recommendations of "tailor-made" training have a particular relevance in these children.

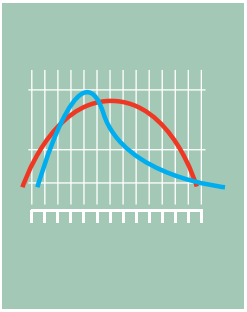
The functional assessment prior to the exercise program is essential to know the initial state of the subject and to be able to adapt the program to their functional capacity. Functional capacity evaluation allows us to objectively measure physical condition, which is the set of physical qualities that enable us to perform everyday tasks.

The exercise at the right dose obtains adaptations in different organs and systems.

Figure 1. (Ergometer, Gases analyzer Vmax, ECG and Pulse Oximeter).

Laboratory of Physiology of exercise at the rehabilitation service in the Children's University Hospital "Niño Jesús" of Madrid.

It is necessary to try to maintain aerobic capacity, in other words, the ability to perform physical activity of low to moderate intensity and for prolong periods; a quality that well developed allows carry out the day-to-day activities better. It is precisely this quality that best relates to the limiting factor of these patients, which is ventilatory restriction (bronchiolar obstruction and the weakness in ventilatory muscles, among others). We also want to evaluate muscle strength and flexibility, which are also useful for performing the activities of everyday life.



Etiological diagnosis of the limitation of the exercise
- · Deconditioned muscle
- · Respiratory limitation
- · Cardiovascular limitation

Prescription of exercise

Exercise Intensity
- · Anaerobic Threshold
- · Below the desaturation of oxygen

Medical diagnosis

· Control on the progression
- · Of the illness
- · On treatment effectiveness

· Transplant indication

Figure 2. Ergoespirometric data obtained in a stress test with metabolic analysis and profits of the PE. *(modified Teoh 2009)*.

The stress test is performed with continuous electro cardiographic registration, controlling saturation of O_2 and analyzing in real time the metabolic variables and ventilatory values that allow us to know the aerobic capacity of the patient in a more realistic way. This is due to the body in motion which can manifest alterations that otherwise go unnoticed; often appearing cardiovascular alterations, such as right ventricular overload because of an increase in pulmonary vascular resistance, which can lead to right ventricular hypertrophy and that could even compromise the left ventricular filling by septal shunt *(Nici et al. 2006)*. In the more developed Cystic Fibrosis patients and therefore with forced expiratory volume in one second (FEV_1) less than 50% of normal, may compromise the stroke volume and cardiac output (amount of blood that the myocardium is able to move). Sometimes this disorder, can lead to diastolic dysfunction of the left ventricle, even at rest *(Todd et al. 2003)*.

More than 90% of the energy that we use throughout the day is supplied by the aerobic metabolism, we get, in essence, from the combination of oxygen with fatty acids and glucose provided by food. When we are at rest, practically all the energy comes from combining oxygen with fatty acids. When we move and depending on the intensity, it begins to intervene the glucose as a nutrient. The result is energy for the muscles, which "we package" in molecules of adenosine triphosphate (ATP) and the production of water and carbon dioxide or anhidric-carbon (CO_2). If we progressively increase the intensity of the workout, we reach an intensity of effort, which is different for each person, this triggers another complementary mechanism from the first, called anaerobic metabolism (anaerobic Glycolysis) to provide energy more quickly. As its name indicates glycolysis (glucose break), splits a monosaccharide of 6 carbons into, producing two molecules of 3 carbons, called lactic acid. This implementation of the anaerobic mechanism is called anaerobic threshold and complements the energy production required to maintain a certain intensity of effort.

Accurately measuring these intensities, helps us to establish the stimulus needed to enable adaptation of the different systems and improve the function of the different organic systems. If we make the analogy with a drug, we could say that these findings help us to know the milligrams of drug, which is effective to achieve the effect we seek.

For that, what we do in our Clinical Exercise Physiology Laboratory consists of precisely that: design a proper physical exercise for children with Cystic Fibrosis that allows us to define the intensity where their anaerobic threshold is and the maximum capture of oxygen that is ultimately the maximum capacity to capture power. These data allow us to adjust the training, taking full advantage of the improvement which this exercise stimulus generates in the different organs and systems.

The stress test enables us to distinguish the cause of an altered response to exercise, as the cause of exercise limitation;

also certainly, it is the most effective tool for the diagnosis of the pathophysiology of the cardiovascular and respiratory systems.

Unlike other diagnostic tests, which usually evaluate an isolated organ, the ergoespirometry during a ST analyzes each one of the systems involved in the physical activity. For example, a test that only apply the electrocardiogram (ECG) during exercise, can tell us nothing more than the response of the myocardium, which limits the sensitivity and specificity of the test. Sometimes children with Cystic Fibrosis may suffer mix cardiopulmonary disorders. This test can be used for the differentiation of the predominance of one system or the other in the limited functional capacity of that patient before deciding treatment *(Weberet, 1997)*.

Figure 3 summarizes the gears between the respiratory system, with the absorption of oxygen and the release into the atmosphere of carbon dioxide (VO_2 and VCO_2), the consumption of oxygen by mitochondria (QO_2) to produce energy and the production of carbon dioxide (QCO_2), as a result.

Figure 3. Perfect gear among organic systems to get the necessary power for proper operation during physical exertion.

Exercise in the right dose manages to improve the VO_2 Peak and the anaerobic threshold, which provides an improvement in day-to-day activities and a better prognosis of the disease.

Naturally, the bloodstream has to increase to accommodate the increasing needs of oxygen (QO_2) of cells, and therefore the amount of blood that comes from the heart must increase also proportionally to these (QO_2). In healthy people, when the intensity of the exercise is constant, the blood flow to the muscles increases more than 5 litres of blood per litre of oxygen consumed. Because 5 litres of arterial blood contains 1 litre of oxygen, it is necessary to increase proportionally the amount of circulating blood for increased oxygen needs, when you boost the workout intensity.

When there is any disorder, as in the case of the cystic fibrosis, in which it is not possible to maintain the increase in obtaining oxygen from the atmosphere (VO_2) proportional to the needs of the mitochondria of the muscles (QO_2), it starts anaerobic metabolism *(anaerobic threshold)* to an intensity of exercise less than what would happen in a healthy person.

The increase in the production of carbon dioxide (QCO_2) without a good ventilatory response acidifies the blood, which limits the ability to continue that effort intensity for anytime. The duration of the session is limited by the respiratory inefficiency.

One of the main parameters that we determine automatically in these tests is the peak oxygen consumption (VO_2 Peak), in other words, it is the maximum amount that the patient is able to consume at the time of maximum intensity of exercise that has been able to endure during the test. It is usually measured in millilitres of oxygen per kilogram per minute ($mL \cdot kg^{-1} \cdot min^{-1}$). This factor, considered as the main predictor of active life, both in healthy people and in patients *(Blair et al. 2004)*, usually evolve negatively over time every year around 2 $mL \cdot kg^{-1} \cdot min^{-1}$ *(Pianosi et al. 2005)*.

In addition, it has been found that people with less than 32 $mL \cdot kg^{-1} \cdot min^{-1}$ of VO_2 Peak might suffer a 60% additional risk of mortality in the next 8 years. However, none of the patients evaluated with VO_2 Peak higher than 45 $mL \cdot kg^{-1} \cdot min^{-1}$ died in the analyzed period of 8 years *(Pianosi et al. 2005)*. There is also a correlation of early mortality, which refers to the pace of deterioration in FEV_1 *(Pianosi et al. 2005)*.

The low VO_2 Peak in patients with FEV_1 is associated with an increased risk of hospitalization for pulmonary exacerbations (Pérez et al. 2014 M).

Our group has extensive experience in the functional evaluation of children with chronic diseases. We are in favour of the use of the treadmill as ergometer with children, except those who present a disability then we discourage it. It is mainly due to the ease of children to adapt to the ergometer and ramp protocols, where the workload is very progressive, simultaneously increase the speed and the inclination of the treadmill, until in most cases reach 90% of the maximum heart rate and/or respiratory quotients >1.09 (San Juan 2008).

Methodology of an exercise stress test (or treadmill test)

The design of an exercise stress test depends on the specific features of each patient.

For children 6 years old and above, and over 120 cm in height, we apply ramp protocols, which are exercises where the intensity is increased and continuous. Starting from an initial speed of between 1 and 3 km·h-1 and a slope of 0.5%, with equivalent increases in speed of 0.1 km·h-1 and the slope of 0.5%, added every 15 seconds. In younger children, the initial speed is 1-2 km·h-1 and 0.5% slope, with increases of 0.1 km·h-1 and slope of 0.25%, added, every 15 seconds.

As we already noted, maximum heart rate is difficult to predict in children with Cystic Fibrosis with the classical equa-

tions depending on age, so it must be obtained during the exercise stress test directly.

In addition to the ECG, we systematically use monitoring of the saturation of oxyhemoglobin during the test, especially in more advanced cases (with FEV1 less than 50% of normal). If significant desaturation is observed (< 80%), there maybe other types of functional tests such as walking for 6 minutes. When the purpose is to design of the training program and with the objective of low rates of saturation (< 80%) both at rest and during the test, we should be training with additional oxygen.

Sometimes appears a persistent cough during the test, which can force us to interrupt the ergometry, although in most cases the test is usually concluded properly.

There are some medications whose effects during the exercise stress test on patients with Cystic Fibrosis should be mentioned. For example, beta-agonist bronchodilators might induce tachycardia. Oral or inhaled corticosteroids reduce the inflammation and bronchospasm in the airway. It has been observed myopathies, diabetes and high blood pressure induced by oral corticosteroids use. Cromoglycate can prevent exercise-induced bronchospasm. The bronchodilator teofilins drugs can also cause tachycardia, tachypnea, or any arrhythmia. Ipratropium, with good bronchodilator effect, can also trigger tachycardia. All these drug-response interactions to the exercise stress test need to be properly evaluated by the team that analyzes the patient.

Strength and Flexibility

Regarding the state of muscle strength in people with Cystic Fibrosis, a decrease of 25% in muscular efficiency has been observed, probably due to the limitation of the mitochondrial activity *(Meer, 1995)*. Loss of muscle protein, which may appear by nutrition disorders, for example derived from pancreatic alterations, tends to be exacerbated by the sedentary lifestyle and the inflammatory phenomenon. The fatigue in

the legs, very characteristic of most of the respiratory diseases, *(Lamhonwah 2010)* is also present here.

Unlike the above, muscle diaphragm usually resists fatigue, due to the overload that it usually supports, though occasionally this high blood supply is available, as a well trained muscle, may extract blood from other muscles involved in the exercise, limiting their activity. *(Nici 2006)*.

The strength exercise improves the quality in all muscular groups involved and it has a positive impact on the development of strength.

The flexibility is not usually an element of physical condition, which alters in Cystic Fibrosis children, although some patients may develop an osteohypertrophy, low bone density and vertebral disorders that will end up limiting said quality *(Boas 1997)*.

The unit of measurement used to estimate muscle force is based on determining the most weight that the patient is able to move or lift during 5 complete repetitions through full range of movement but not with 6 (5 repetitions maximum or 5-RM). Naturally, it is different for each muscular group. We chose the most representative: leg press, leg extension, leg curls, pull downs, seated rowing, and abs. We complement these tests with exercises that simulate everyday tasks, such as climbing and descending stairs, getting up from a chair, walking fast; using the corresponding test for each muscle group, which assesses its strength and speed of execution.

Detail of the tests for the evaluation of strength, up and down stairs.

Go Step by step for a good physical condition which guarantees us a better quality of life.

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05

HOW TO DO AND PLAN EXERCISE AT THE RIGHT DOSE?

Elena Santana Sosa
Doctor in Sport and Physical Activity
Children's University Hospital "Niño Jesús" of Madrid.
Aladina Foundation
E-mail: santana.sosa.e@gmail.com

How to make and plan the exercise at the right dose?

As explained in the preceding chapters, there are a large number of studies that have investigated the potential of exercise programs and their benefits in Cystic Fibrosis.

This chapter will present guidelines of how to make a progressive safe and effective training based on scientific evidence.

Before beginning

Before starting any type of sport exercise program, we have to be clear about what are our goals and needs (lose weight, gain muscle mass, improve our physical condition in general...).

Once the objective is clear, we have to know in what condition we are.

Beginner	Intermediate	Advanced
Does not train or just started the program	Training between 2 and 6 months	Training over 6 months

Then we have to set a frequency of training (1-2 days/week; 3-4 days/week), then select the exercises and the order they are carried out.

In order to meet these objectives it is important to personalize the training program, number of repetitions, weight used, as well as the volume and rest periods.

Variables for designing a training program
1.-Analyze objectives and needs
2.-State of physical condition
3.-Frequency of training
4.-Select the exercises and the order they are executed in
5.-Training load and repetitions
6.-Volume
7.-Rest periods

How to calculate the workout intensity?

The intensity of the exercise indicates how hard the workout is. If we do low intensity exercises we will not achieve our objective, if on the other hand if we do exercises, which are too intense, it will be difficult to maintain the routine and it will increase the chances of suffering injury.

There are several ways to control the intensity of aerobic exercise:

The best way is through an exercise stress test (Chapter 4). If that is not possible, we can estimate it by means of the maximum heart rate (MHR) with the help of a heart rate monitor (watch which records heart rate), and the third option is via the speech test.

To calculate strength training load:

For a correct workout of strength, you must know what is your maximum load in each exercise in order to program your workout. This value is known as one maximum repetition or 1 RM. The 1 RM test is a very intense and not recommended for beginners or children, so we recommend doing a 5 RM test.

How do I do it?

We know that moderate to vigorous exercise is as effective as postural drainage and percussion that protects against the deterioration of lung function and has obvious effects of well-being in this pathology.

We have also found that combined aerobics and strength exercise programs entails positive effects on functional capacity and muscle strength.

Therefore, our recommendation is as follows:

According to our physical condition and our marked objectives, we should do a routine of 30 minutes of aerobic exercise and 20-25 minutes of **strength exercises**.

How to know your ideal HR		
		Example
Maximum HR	220 - age	220 - 25 = 195
Moderate activity	The level of 60% is HR x 0.60 = ¿? beats per minute (bpm) The level of 70% is HR x 0.70 = ¿? bpm.	The level of 60% is 195 x 0.60 = 117 bpm. The level of 70% is 195 x 0.70 = 136 bpm. The ideal heart rate would be between 117 and 136 bpm.
Vigorous Activity	The level of 70% is HR x 0.70 = ¿? bpm. The level of 80% is HR x 0.80 = ¿?bpm.	The level of 70% is 195 x 0.70 = 136 bpm. The level of 80% is 195 x 0.80 = 156 bpm. The ideal heart rate is between 136 and 156 bpm.

Strength training:

As we recommended in the previous point, the ideal is to assess each muscle group that you want to work, through the 5 RM test.

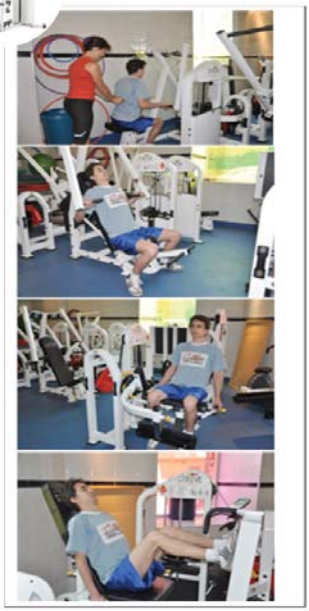
The result of this test gives us a value of 88% of our maximum strength. So to estimate 100% of the force will use the Brzycki formula.

How much weight should I lift on each exercise machine?	
Brzycki formula	1RM=weight lifted / (1.0278-(0.0278 x Number of repetitions)).
Example: Leg Press 70kg	1RM= 70 kg / (1.0278-(0.0278 x 5)) 1RM= 70kg / 0.88 1RM= 79.5 Kg

Practical applications:

Aerobic exercise

Strength machines



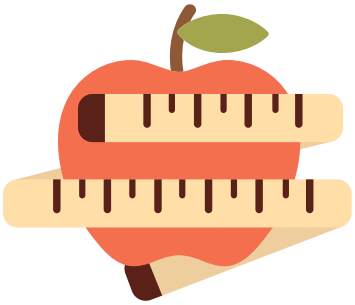
Alternative options

Example of a mixed-type circuit where aerobic resistance is being worked, trying to keep an intensity of 75% of the theoretical maximum heart rate, with minimum intervals of 10-12 minutes up to the full dose of 30 min, interspersed with **strength exercises** for large muscles groups in each one of the machines.

Example of strength **circuit for upper body training without machines**, using rubber bands of different strength and weight of the subject.

Examples of **upper body stretching**.

GUIDELINES FOR PATIENTS WHO ALSO HAVE DIABETES



Margarita Pérez Ruiz
Doctor in Medicine and Specialist in Sports Medicine
Professor of Exercise Physiology at the European University of Madrid
E-mail: margarita.perez@universidadeuropea.es

Verónica Sanz Santiago
Doctor in Medicine
Specialist in Pediatric Neumology.

Guidelines for the patient who also suffers from diabetes

There is evidence that physical exercise has an important health benefit and that sedentary life habits are associated with an increase in many chronic diseases such as diabetes mellitus type 2 (DM2). The exercise is a therapeutic component essential in diabetes management, although difficult to comply with it, can be used to promote health and quality of life for these patients. It is considered that moderate levels of physical exercise produce considerable benefits, especially in the DM2.

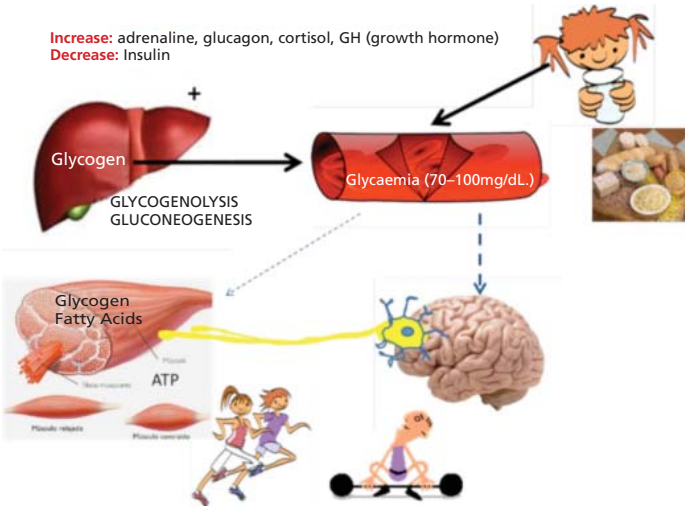
In CF, endocrine pancreas affectation causes a decrease in the secretion of insulin, which leads to an alteration in the metabolism of carbohydrates and that may evolve into diabetes. Abnormal function of the channels of the chloride ion in the CF involve viscous secretions that cause obstruction of the exocrine pancreas with progressive fibrosis and adipose infiltration.

Exercise at the right dose corrects resistance to insulin (type 2 diabetes) but also helps to decrease the dose of insulin necessary for the treatment of type 1 diabetes.

In addition, exercise acts as a preventive tool for the reduction of cardiovascular ailments that in the long term may be associated with diabetes type 1 and 2.

Metabolic response to exercise and Glycaemia control

When exercising, muscle glucose energy demand boosts the stimulation of a number of hormones that trigger the glycogenolysis and gluconeogenesis in order to maintain liver glucose production and, therefore, basal glycaemia at optimal values for the functioning of all organs and systems and a decrease of the levels of insulin. Plasma glucose levels remain constant, since the increase in glucose uptake in the contracted muscles is compensated by an increase in the hepatic production of glucose (glycogenolysis and gluconeogenesis). When the exercise is long lasting the maintenance of glucose can also be obtained via drinks and supplements containing carbohydrates.



When you start the exercise program alert your endocrine and your sport physician, because they must adjust the time of day, place and the dose of the insulin injection needed.

BENEFITS OF EXERCISE

- **On the glycemic control**
- **On total mortality rate**
- **On cardiovascular risk**
  - – Hypertension
  - – Dyslipidemia
  - – Insulin resistance
  - – Cardiovascular disease
  - – Obesity
  - – Fibrinolytic system
- **Psychological**
  - - Decreases stress response
  - - Reduction of anxiety
  - - Decreases depression
  - - Improves self-esteem
- **Economical**
- **Prevention of diabetes**

However, in diabetes mellitus type 1 (DM1) and in DM2, with insulinopenia (Hypoinsulinism) and insulin therapy, although it remains the response of counterregulatory hormones, no changes occur in levels of insulinemia. In this group of patients, exercise can cause a decrease, an increase or a maintenance of glucose level, depending on the circumstances in which it is performed and if it has been carried out or not a prior adjustment of insulin doses to the new lifestyle.

Physical exercise leads to a lowering of blood glucose in patients with type 2 Diabetes and an increase of the sensibility to the insulin that lasts 12-72 hours after exercise. The effect of regular physical exercise contribute to improved glycaemia control in the long term for patients with diabetes mellitus type 2, particularly in those in which predominates insulin resistance.

Glycosylated hemoglobin is a laboratory test used in diabetes to know if the glycemic control carried out by the patient has been correct for the past two months. It also helps us as a procedure to find out if your blood sugar is maintained at correct levels for exercising without worrying about a rise in sugar levels in blood, given that vigorous exercise stimulate glycogenolysis and could cause an increase in blood glucose levels.

Less than 7% glycosylated hemoglobin, reduces considerably the risk of micro and macro vascular complications.

We must know that the intensity of the exercise performed influence more in reducing HbA1c than the volume of exercise, so that once a good physical condition and a knowledge of the glycemic response to exercise is achieved, you should try to work intensely in short periods of time.

Exercise has beneficial effects on HbA1c regardless of body weight.

However, these beneficial changes generally deteriorate after 72 hours of the last exercise session. The impact of exercise on insulin sensitivity stays between 24 and 72 hours, depending on factors such as duration and intensity of the exercise. Hence, there is a recommendation for daily physical exercise or, at least, three days a week, avoid being more than two consecutive days without doing any.

In patients with type 1 diabetes or those with type 2 diabetes in which predominates the insulin deficiency, the effects of physical exercise on blood glucose are more variable and may cause decrease, increase, or no alteration.

This is due to that the physiological hormone changes that occur in people not diabetic during exercise, do not occur due to lack of automatic adjustment of insulin.

Therefore, the changes in blood glucose with exercise will depend primarily on existing insulin levels. In these patients, although physical exercise reduces insulin requirements, it does not usually improve long-term glycemic control. Hence, exercise may not be intended to meet this goal, but for many other reasons related to health and, especially in children and young people.

Exercise produces glycemic control in DM type 2, but not in DM type 1.

Physical exercise reduces the requirement of insulin in the DM type 1.

It has been shown an inverse correlation between physical activity and mortality from any cause in the general population and DM2. Inactivity is a predictive factor independent of total mortality in these patients and even the risk of death increases with less-favourable Glycemic status, the beneficial effect of exercise is greater than the expected by the decrease of the glycaemia and it is also independent of body weight.

The exercise also influences changes in the distribution of body fat.

Doing physical exercise, as in non-diabetic people, has a positive impact on cardiovascular mortality. This protective effect is independent of factors such as sex, age, body mass index and time suffered with diabetes. Hypertension is present in up to 60% of the patients with DM2. In non-diabetic

people, physical exercise has an independent effect on the reduction of blood pressure, with a decrease in systolic blood pressure and in diastolic blood pressures.

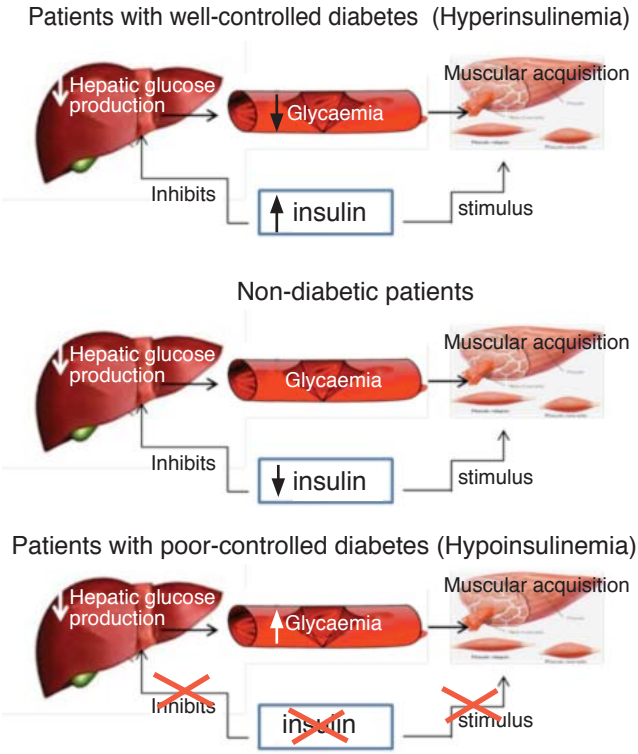
Physical exercise and glycemic changes in patients treated with insulin.

HYPOGLYCEMIA

Under a of good glycemic control and at times of day when insulin is higher, if the insulin levels do not diminish, or increase Carbohydrates (CH) intake before or during physical exercise, there is a risk of hypoglycemia. This is due to the relative hyperinsulinemia inhibits hepatic glucose production and increases its peripheral acquisition, so levels of glucose will decrease and may result in hypoglycemia.

Higher risk situations correspond to those when exercise is carried out coinciding with the peak of the administered insulin; for example, after meals in the guidelines with multiple doses, or during the morning and after dinner in the guidelines with 2 doses of insulin NPH or intermediate. The risk of hypoglycemia will be higher when the physical exercise is more intense and prolonged.

Glucose Hemeostasis during exercise



HYPERGLYCEMIA

When insulin levels are too low (situation of poor metabolic and functional control due to the dose of insulin), exercise causes a greater increase in the production of glucose (release of downregulator hormones and low insulin levels) and its utilization by muscle is decreased (deficit of insulin), therefore instead of hypoglycaemia, hyperglycaemia will appear and even Ketosis, where lack of insulin leads to metabolize fatty acids as an alternative energy source.

Therefore, it is important to monitor blood glucose prior to doing exercise and if this is greater than 250-300 mg/dL, determine the Ketonemia/Ketonuria, as if it were positive, physical exercise would be counterproductive until glycemic control is restored, to avoid a situation of greater Ketoacidosis.

Despite a good glycemic control prior to the exercise, Hyperglycemia may appear after a short, very intense or violent physical activity, with anaerobic characteristics, or after a competition where physical and/or psychological stress leads to an important peak of counterregulatory hormones secretion and the production of hepatic glucose, which overcomes the intake.

In these cases, if insulin is enough and therefore Hyperglycemia is not accompanied by ketosis, it can be solved by the intake of CH and/or increasing the dose of insulin before exercise. Another cause of Hyperglycemia after exercise is the excessive intake of dietary supplements before or during this, carried out frequently to avoid hypoglycemia.

Exercise prescription:

We must try to prevent changes in the Glycemia. But how?

1. Reducing the insulin dose if we know we are going to do exercise.
2. Taking a supplement of carbohydrates when we are going to perform the programmed exercise.

What should I choose to do?

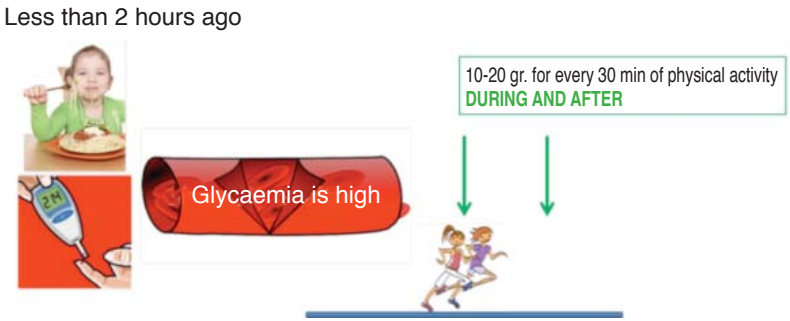
If exercise is not set or programmed: take extra carbohydrates.

If exercise was set or programmed: I can choose between taking CH or lowering the dose of insulin.

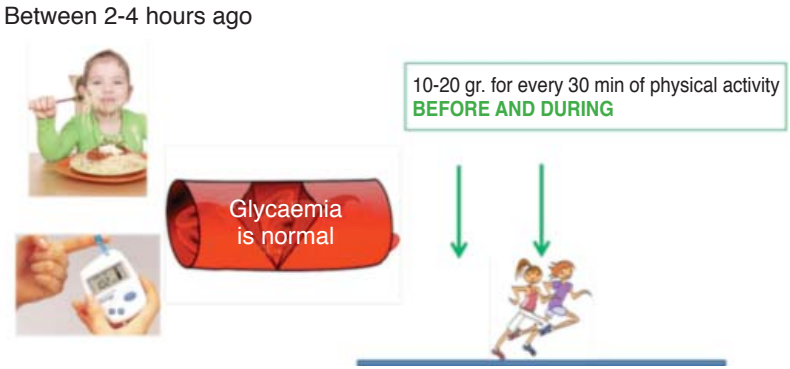
If exercise is not set or programmed

In these situations the dose of insulin which is administrated, cannot be modified, so the only option to avoid hypoglycaemia in the majority of cases, will be taking a supplement of CH (usually about 10-20 g. is enough for every 30 minutes of physical activity).

If the prior glucose level is high or more than 2 hours has passed since the last meal: The CH supplement must be taken during and after exercise.



If the prior glucose level is normal or more than 2 hours has passed since the last meal: the CH supplement must be taken before and during exercise.



If after exercising, the level of glycaemia is low, on other occasions you should increase the amount of CH supplements taken.

If after exercising, the level of glycaemia is high, on other occasions you should decrease the amount CH supplements taken.

Programmed or set exercise

In these cases, a prior exercise strategy can be set to avoid a hypoglycemia. It is not clear if it is better to lower insulin doses or to take an additional CH supplement. It will depend on patient preferences on how long the exercise has been carried out, but remember that a hyperinsulinemia condition will inhibit the endogenous production of glucose and muscle performance will be lower. On the other hand, a situation of hypoinsulinemia will prevent muscle glucose uptake and muscle performance also descends.

What changes when I am diabetic and I do exercise?

· The time of doing exercise, adapt yourself to the blood glucose levels (100 - 250 mg/dl).
· Measure blood glucose before, during and after exercise.
· CH supplement should not be forgotten.
· Adjust the insulin dosage.
· Avoid injecting insulin in areas of the body which you are going to exercise.
· When using an analogue of rapid insulin, exercise should not be performed within 2 hours after its administration (risk of hypoglycaemia).
· Find the most appropriate schedule according to the insulin administration and the insulin type. The ideal is that when you start exercising this insulin must be at low concentrations.
· Learn and get diabetes self-monitoring.

What I should not change even if I am diabetic?

· The need to do exercise.
· The recommended dose (it is preferable intensity instead of volume when you are in good physical condition).
· CH supplement is needed when doing exercise.
· Good hydration.

What do I need to know?

· The effect of exercise on blood glucose may persist for 12-24 hours after it has been done.
· Adjust the extra intake of CH or reduce insulin dosage.

To prevent decreases of glucose in blood:
· Carry out more tests of blood glucose in order to analyze its behaviour and act accordingly.
· Take more CH supplements with respect to days that exercise is not performed.
· Possible adjustments of insulin treatment.

Precautions
· Carry sweetened drink, as the risk of low glucose levels is high in the first sessions.
· Better to exercise in the morning.

Do not exercise if...
High glucose levels 300 mg/dl and/or Ketonuria is present.

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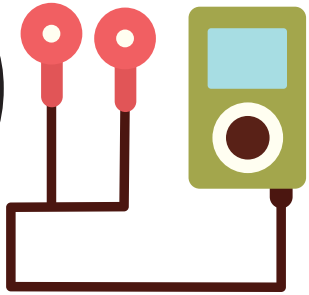
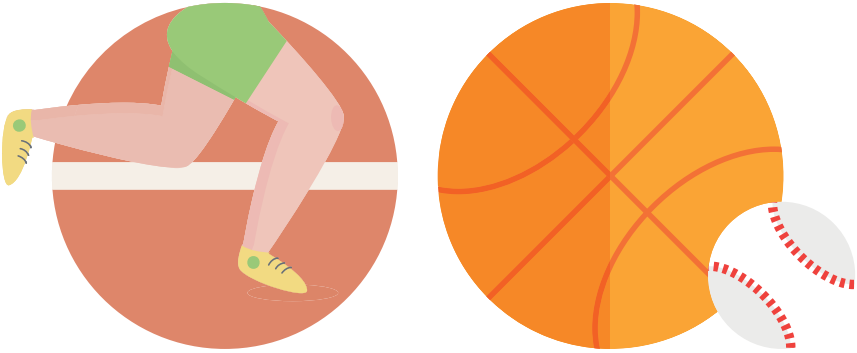


FREQUENTLY ASKED QUESTIONS RAISED BY ALL THOSE WHO WANT TO START EXERCISING.

Margarita Pérez Ruiz
Doctor in Medicine and Specialist in Sports Medicine
Professor of Exercise Physiology at the European University of Madrid
European University of Madrid
E-mail: margarita.perez@universidadeuropea.es

José Ramón Villa
Doctor in Medicine and Specialist in Pediatric Neumology.
Head of Neumology service at Children's University Hospital "Niño Jesús" of Madrid (HUNJ)



What do you need to start doing a sport?

Before starting to play a sport you need, just like anyone else without your disease, to achieve a good physical condition.

Exercise at the right dose and adapted to your initial fitness level, will let you slowly reach a proper level of physical condition, so you can participate comfortably in a sport. The component of physical fitness which has been most frequently studied in its association with health is the aerobic power or, as measured in the laboratory of physiology, the maximum consumption of oxygen.

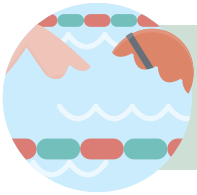
This attribute is also called cardiovascular, respiratory or resistance capacity. Getting physically fit will let you easily develop the activities of everyday life and will allow you to participate in the sport that you like.

Sport also involves rules, technique, tactics and teamwork. All of these are important to better face current and future working life.

Which sport suits you best?

Without doubt, the one that you like most, because the important thing is that once started a sport, at the same time helps to maintain your physical fitness. To achieve that purpose you need to adhere to it that means doing it at least weekly.

Being consistent is the basis of success.



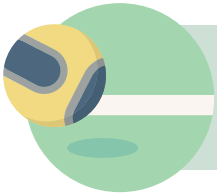
SWIMMING

If you choose swimming, you should know that although it is positive for training your respiratory muscles and therefore your lung capacity, chlorine in swimming pools can irritate the respiratory mucosa and generate inflammation in it, this would not be entirely appropriate. You should know that there are ways to train respiratory muscles in the dry. How? If you mention it to your physical therapist, he knows how to adapt a good respiratory training program with apparatus that generates some breathing resistance.

The choice of a safe pool for your children will allow them to achieve the benefits of swimming without the threat of bacterial infection.

Swimming is not a weight bearing exercise, so it is not the most appropriate sport to prevent osteopenia. Young people do not have osteoporosis but they need to build enough bone density for when the time of physiological loss, that occurs in all subjects at a certain age. If you are now young, you need to do sports that load your weight, thus contribute to achieving a greater bone density to face the physiological loss that occurs in any subject as age advances.

In addition, you should know that continued glucocorticoids intake may predispose you as an adult to suffer osteopenia or osteoporosis earlier.



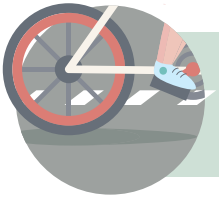
SOCCER

If you choose football, you should know that it is a very positive team sport to promote cooperation, values of coexistence and you also work body coordination and changing direction while running. That stimulates depositing of calcium in the bones, which helps you to acquire bone density. Compensates in some way the negative effect of glucocorticoids if it is a drug that you need to take.

If you are already an adult and you like this sport, take care with your workouts to minimise the risk of injury and with it you will slow down bone density loss. If you already know that you suffer from osteoporosis, this is not the best sport, since the risk of falling is high and the bone could be broken.

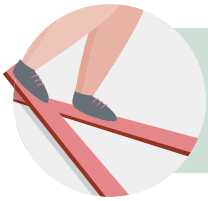
BASKETBALL

If you choose basketball, you can add to the above jumping with your body weight that asks more of the bone, so the bone density you get, if you play it before puberty, is very appropriate.



CYCLING

If you choose cycling, you avoid overloading your joints, since the work of the bicycle requires a concentric muscle contraction. With this, you will not improve your bone density but if you are already an adult, may be it is enough.



SKIING

If you choose skiing, you should avoid doing it in high-altitude mountains. From 3000-4000 meters high, the atmosphere has a low oxygen pressure, causing hyperventilation to compensate for the atmosphere. As we do not stay the necessary time to acclimatise, the body is unable to adapt, which can cause desaturation, hypoxia and right heart failure.

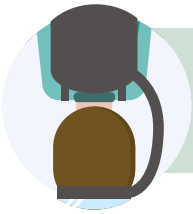
DANCING

If you choose dancing, it is a good sport, which also helps to improve your coordination. Perhaps, in sometimes during adolescence, is the best way to keep your physical condition.

Can I do sport in any public sports facilities?

If the gym is well ventilated and with appropriate hygiene measures, the program is tailored to each user and directed by a professional who knows your pathology, then you do not have any problem. But you should know that it is not recommended to stay indoors with other people who have CF.

In addition, if you are infected with Burkholderia cepacia, MRSA and other contagious and multidrug-resistant microorganisms, you can follow the exercise program, but avoiding contact with peers who have your disease but have not been colonized. (Avoid cross-infection).

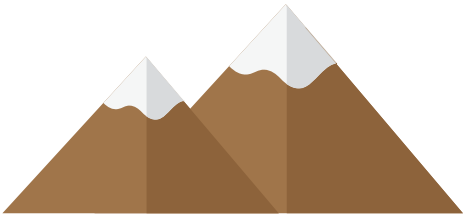


DIVING

Can I do scuba diving?

You should know that you increase the risk of pneumothorax, especially in advanced CF. You should avoid exercises that involve sudden intrapulmonary pressure changes.

Can I go up to high altitudes?



High altitudes do not have an appropriate atmosphere for humans; we achieve acclimatisation to it if we ascend slowly and if we stay in camps long enough (2-3 days). Time in which the respiratory system responds hyperventilating and thus manages to compensate for low atmospheric pressure of oxygen and kidneys compensate for alkalosis caused by hyperventilation removing bicarbonate and water through the urine. We need to know about this for drinking water even if we are not thirsty.

It is not an atmosphere suitable for people with CF, but if ever faced with that situation, it is important to monitor oxygen saturation and the symptoms of hypoxia, such as headache and if they should occur, it is necessary to descend to a lower altitude.

Pool humidity can encourage Pseudomonas infection?

Pseudomonas aeruginosa is the microorganism infecting bronchi of people with CF more frequently; nevertheless it is rare in people who do not have this disease. People with CF have a special predisposition to

attract this pathogen to their bronchi and once it is acquired, it is difficult to eradicate. Pseudomonas aeruginosa is a bacteria widely distributed in nature. It can be found in natural environments (soil, water), in aqueous reservoirs (sinks and faucets in paediatric rooms), drains, toilets, cleaning supplies, home nebulizers, peak flow meters and toys. Also in swimming pools, hot tubs and dental equipment, even in plants and animals. And it is also found on the hands of people with CF or medical personnel. Hence the need to maintain strict hygiene, especially with a correct hand washing.

Hot water and soap eliminate most of the bacteria and correct hand washing prevents the spread in almost 100% of infections. Therefore the importance of proper cleaning and disinfection of nebulizer equipments.

According to a 2009 report on "Reviews of Environmental Contamination and Toxicology", the Pseudomona aeruginosa is a naturally occurring pathogen and, sometimes, is located in whirlpool baths, water slides and pools that have inadequate levels of chlorine. Rivers and lakes are also often fertile ground for this bacteria. This bacteria is susceptible to chlorine. If your child with CF is going to swim, make sure that pool is well maintained, having the right pH and chlorine levels. A swimming pool which is maintained regularly must be secure so your child can swim.

Check... the pool and its surroundings. What should be noted?

- Clean and clear water. You should be able to see clearly any painted stripes and the bottom of the pool.
- Make sure the sides of the pool are smooth; tiles should not be sticky or slippery.
- That there are no odours, a well-chlorinated swimming pool has little odour. A strong chemical odour indicates a maintenance problem.
- The pool equipment is functioning; pool filtration systems and pumps make noise and you should hear them working.

Can I do strength exercises?

Not only you can, but also you should do it, because it is essential for children's development. Before puberty, the best games are those, which involve strength, as this is a very important quality to develop. If I am adult, it is best to do strength training with exercise equipment helped by a professional who set us the recommendations according to initial muscular capacity and its progression. The muscle is a locomotive and endocrine organ that when working at the correct dose manages to release anti-inflammatory cytokines counteracting the general inflammatory status.

The National Strength and Conditioning Association (NSCA) published in 1985 the first report on the importance of strength training in young people and we recommend the most appropriate protocol in terms of the number, series, repetitions and loads for each functional unit of muscle, according to the pubertal development stage in the child.

Can I do exercise in high humidity and hot environments?

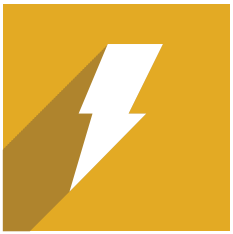
No one should perform exercise in these circumstances because they increase the risk of heat stroke. For you, when you have CF, it is a factor that worsens the risk of alteration of the electrolyte balance, hyponatremia and dehydration, so it is not a recommended environment but if you become exposed to this climate, remember to replenish salts during the exercise, by drinking water and isotonic drinks every 15 minutes.

Should I avoid to get becoming exhausted or tired?

It is not bad to feel tired after exercise, fatigue is a positive indicator that we must listen to and we must give ourselves enough active recovery time before starting again the next set of exercises. As you acquire better physical condition, you will have that feeling of tiredness later, since the body has adapted to different stimuli, but you should give some time to achieve this.

What is known is that any exercise, as little as it may be, is better than no exercise at all. Numerous studies have shown that workouts in intervals are better tolerated by people with respiratory problems with regard to resistance exercises.

What is muscle stiffness?

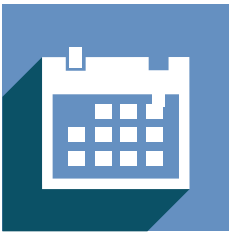


It is the colloquial name for the muscle pain called delayed onset muscle soreness (DOMS) or muscle pain post-effort late onset accompanied by a muscle inflammation.

When the muscle suffers stiffness, it needs rest, what it gets back is adaptation and improvement. Then you will notice that you need to increase your training load to get it again. It is recommended to perform one session of exercise a month that causes you stiffness in the different muscle chains to improve the muscular system.

It is advisable to warm-up, as well as a progressive increase in the level of training, starting with gentle up to the most intense exercises. Thus, the muscle fibers are prepared for a situation of effort.

How can I generate exercise adherence and get to do it every day?



Before puberty, you only get adherence to exercise to develop the physical qualities and improve physical fitness, if you incorporate playing games into the exercise programs. It is very important to involve the family, since children are reflections of their parents. Therefore, parents should exercise because, although they do not suffer from CF, they also benefit from it. In a healthy lifestyle, they have to get into these habits before the age of 12.

After puberty, we will only achieve that the teenager performs exercise if we develop the habit in childhood and we practice what we preach. If they are in good physical condition, it is best to do team sports, so we ensure that their friends are also committed to exercise and do it.

As an adult if you have never done exercise it is the time to start, start exercising very slowly, at the dose that fits your physical condition, this is the way forward and reach your objectives. You will get the adherence to exercise if you feel healthier every day and you will have less pulmonary exacerbations (remember that it is one of the effects of the exercise).

More than 745 scientific articles endorse the need for exercise as therapy in CF.

Make exercise part of
your lifestyle,
so it will be easier for you
to comply with this treatment,
which is as important
as any other.

Complications associated with the CF and special cares.

When you have complications, exercise must be adapted to the evolution of the disease, but you should always know that rest is the least appropriate in almost all circumstances.

If you have pulmonary exacerbation, you have to wait until the severe episode improves and as soon as the acute symptoms disappears, resumes your workouts, checking oxygen saturation and heart rate. It is normal that you get tired early and your physical condition worsens, but after only 4 weeks, you will be the same as before.

If you have had a pneumothorax, resume the exercise gradually, avoid doing upper body strength exercises, do not consider diving.

If you suffer from malnutrition, keep on exercising, since with it you continue maintaining the muscle mass that otherwise you would lose.

If you have hemoptysis, do not perform sudden exercises until it is resolved, gradually resumes the exercise and avoid uncontrolled and vigorous coughs.

If you have liver disease, avoid contact sports (boxing, rugby, wrestling...) in cases of hepatosplenomegaly and be extremely careful with the intensity of the exercise and strength work if you have bleeding esophageal varices.

If you have cardiac impairment, do exercise of moderate intensity, controlling hypoxia and assessing the need for supplemental oxygen.

If you suffer from exercise-induced bronchospasm, do longer warm-ups and cool downs, exercise at the right intensity to keep breathing through the nose.

If you need to breathe through the mouth, try breathing through a scarf to warm the air up before it gets to your

lungs. Carry the inhaled bronchodilator medication in case you have to use it when you finish the exercise or after.

If you have had a lung transplant, restart the exercise as soon as the specialist advice you, begins with flexibility and muscle strengthening exercises, soon you will have back at your physical condition.

EXPERIENCES

My name is Breixo Castro, I have Cystic Fibrosis and I am a student of Physical Activity and Sports Science, but above all, I am a sportsman and I am happy.

My relationship with the sport began as a child, instilled in me by my parents. At school I played on the football team, I went to judo classes and I have always done sports like all my friends, without that my illness being an impediment.

Every three months I had to go to the hospital, to have medical check-ups and tests and I had my relapses as anyone else, but in general, I can say that I am a lucky. And today, at 20 years old, I can assure you that it was and still is thanks to sport.

In the hospital everybody told me that it was very important the physiotherapy and doing a lot of sport and I am aware that they were absolutely right. The sport, in my particular case judo and soccer were more important than any drugs or inhalers. Not only for the tangible benefits when breathing, but because it helped me to deal with the disease much better and most of the time, forget about it.

In addition, over the years I have noticed that in periods when I have done more exercise the hospital tests were better and vice versa. If I had to give one piece of advice, I

would say that everyone should choose the sport they like most, as much as possible and do not do it because you have to but because you enjoy it, knowing that many times it will require willpower and effort, because it is the greatest favour you can do for yourself.

Breixo Castro La Franke
breixocl@gmail.com

Ulises Mateo who also participated actively in the exercise program that began at the Children's University Hospital "Niño Jesús" of Madrid, in 2010.

Few years ago, I was able to participate in a training project and sincerely, it has helped me a lot.

I did cardio and strength work and the improvements were remarkable. They were easy exercises, since they required of simple machines, such as an elastic band, a mattress or even without equipment but I also enjoyed the facilities of the gym.

Also it is worth mentioning, the help of the coaches who explained to me very well how to do each of the exercises and to change the postures we make when doing them which could be quite harmful in the long term (e.g. the curvature of the back).

Clinically, the results were very positive and I managed to keep a good condition.

In conclusion, my experience with the exercise program has been satisfactory. I was able to learn new forms of physical workouts, which are comfortable to put into practice.

Ulises Mateo

07
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Intrahospital Weight and Aerobic Training in Children with Cystic Fibrosis: A Randomized Controlled Trial

ELENA SANTANA SOSA[1], IRIS F. GROENEVELD[2], LAURA GONZALEZ-SAIZ[1], LUIS M. LÓPEZ-MOJARES[1], JOSÉ R. VILLA-ASENSI[3], MARÍA I. BARRIO GONZALEZ[3], STEVEN J. FLECK[4], MARGARITA PÉREZ[1], and ALEJANDRO LUCIA[1]

[1]European University of Madrid, Madrid, SPAIN; [2]Academic Medical Center, University of Amsterdam, Amsterdam, THE NETHERLANDS; [3]Department of Pneumology, Hospital Infantil Universitario Niño Jesus, Madrid, SPAIN; and [4]Sport Science Department, Colorado College, Colorado Springs, CO

ABSTRACT

SOSA, E. S., I. F. GROENEVELD, L. GONZALEZ-SAIZ, L. M. LÓPEZ-MOJARES, J. R. VILLA-ASENSI, M. I. BARRIO GONZALEZ, S. J. FLECK, M. PÉREZ, and A. LUCIA. Intrahospital Weight and Aerobic Training in Children with Cystic Fibrosis: A Randomized Controlled Trial. *Med. Sci. Sports Exerc.*, Vol. 44, No. 1, pp. 2–11, 2012. **Purpose**: The purpose of our study was to assess the effects of an 8-wk intrahospital combined circuit weight and aerobic training program performed by children with cystic fibrosis (of low–moderate severity and stable clinical condition) on the following outcomes: cardiorespiratory fitness ($\dot{V}O_{2peak}$) and muscle strength (five-repetition maximum (5RM) bench press, 5RM leg press, and 5RM seated row) (primary outcomes) and pulmonary function (forced vital capacity, forced expiratory volume in 1 s), weight, body composition, functional mobility (Timed Up and Down Stairs and 3-m Timed Up and Go tests), and quality of life (secondary outcomes). We also determined the effects of a detraining period (4 wk) on the aforementioned outcomes. **Methods**: We performed a randomized controlled trial design. Eleven participants in each group (controls: 7 boys, age = 11 ± 3 yr, body mass index = 17.2 ± 0.8 kg·m^{-2} (mean ± SEM); intervention: 6 boys, age = 10 ± 2 yr, body mass index = 18.4 ± 1.0 kg·m^{-2}) started the study. **Results**: Adherence to training averaged 95.1% ± 7.4%. We observed a significant group × time interaction effect ($P = 0.036$) for $\dot{V}O_{2peak}$. In the intervention group, $\dot{V}O_{2peak}$ significantly increased with training by 3.9 mL·kg^{-1}·min^{-1} (95% confidence interval = 1.8–6.1 mL·kg^{-1}·min^{-1}, $P = 0.002$), whereas it decreased during the detraining period (−3.4 mL·kg^{-1}·min^{-1}, 95% confidence interval = −5.7 to −1.7 mL·kg^{-1}·min^{-1}, $P = 0.001$). In contrast, no significant changes were observed during the study period within the control group. Although significant improvements were also observed after training for all 5RM strength tests ($P < 0.001$ for the interaction effect), the training improvements were not significantly decreased after the detraining period in the intervention group (all $P > 0.1$ for after training vs detraining). We found no significant training benefits in any of the secondary outcomes. **Conclusions**: A short-term combined circuit weight and aerobic training program performed in a hospital setting induces significant benefits in the cardiorespiratory fitness and muscle strength of children with cystic fibrosis. **Key Words:** PEDIATRIC PATIENTS, EXERCISE, $\dot{V}O_{2PEAK}$, MUSCLE STRENGTH

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Health-related quality of life of Spanish children with cystic fibrosis

Iris F. Groeneveld · Elena S. Sosa · Margarita Pérez · Carmen Fiuza-Luces · Laura Gonzalez-Saiz · Cristian Gallardo · Luis M. López-Mojares · Jonatan R. Ruiz · Alejandro Lucia

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Abstract

Purpose To investigate (1) the contributions of sex, age, nutritional status- and physical-fitness-related variables on health-related quality of life (HRQOL) in Spanish children with cystic fibrosis, and (2) the agreement on HRQOL between children and their parents.

Methods In 28 children aged 6–17 years, body mass index percentile, percentage body fat, physical activity, pulmonary function, cardiorespiratory fitness, functional mobility, and dynamic muscle strength were determined using objective measures. HRQOL was measured using the revised version of the cystic fibrosis questionnaire. Simple and multiple linear regression analyses were performed to determine the variables associated with HRQOL. To assess the agreement on HRQOL between children and parents, intra-class correlation coefficients (ICCs) were calculated.

Results Girls reported worse emotional functioning, a higher treatment burden, and more respiratory problems than boys. Greater functional mobility appeared associated with a less favourable body image and more eating

Pediatric Pulmonology

Aerobic Fitness Is Associated With Lower Risk of Hospitalization in Children With Cystic Fibrosis

Margarita Pérez, MD, PhD,[1] Iris F. Groeneveld, PhD,[2] Elena Santana-Sosa, PhD,[1] Carmen Fiuza-Luces, MSc,[1] Laura Gonzalez-Saiz, MSc,[1] José R. Villa-Asensi, MD, PhD,[3] Luis M. López-Mojares, MD, PhD,[1] Margarita Rubio, MD, PhD,[1] and Alejandro Lucia, MD, PhD[1,4]*

Summary. Background: Children with cystic fibrosis (CF) often have to be hospitalized because of acute exacerbation of their respiratory symptoms. Given the fact that improved peak oxygen uptake (VO_{2peak}) is positively associated with lung function and overall health in children with CF, this study examined the association between VO_{2peak} and the need for hospitalization in a cohort of pediatric CF patients. Methods: In a 3-year study, 77 CF children with mild-to-moderately severe CF (forced expiratory volume in 1 sec [FEV_1] $\geq$ 50%) underwent a maximal exercise test to determine VO_{2peak}. Anthropometric, lung function and muscle strength measurements were also conducted and dates of hospitalization were recorded for the study period. Associations were then determined between the variables recorded and hospitalization by univariate and multivariate Cox proportional hazards regression analyses. Results: VO_{2peak} was 38.6 ± 6.7 ml kg^{-1} min^{-1} for boys and 31.9 ± 6.9 ml kg^{-1} min^{-1} for girls. In multivariate analyses, VO_{2peak} was the only variable significantly associated with time to hospitalization (hazard ratio 0.91, $P = 0.03$). Conclusion: A significant association was detected between greater aerobic fitness, and lower risk of hospitalization. Because hospitalization due to respiratory exacerbation is a powerful prognostic factor, our findings provide further support for the importance of aerobic fitness evaluation in the management of children with mild-to-moderately severe CF. Pediatr Pulmonol.

Key words: cystic fibrosis; peak oxygen uptake; hospitalization; children; fitness.

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Original Article

Levels of moderate–vigorous physical activity are low in Spanish children with cystic fibrosis: A comparison with healthy controls

Susana Aznar[a], Cristian Gallardo[a], Carmen Fiuza-Luces[b], Elena Santana-Sosa[b], Luis M. López-Mojares[b], Alfredo Santalla[c], Gabriel Rodríguez-Romo[d], Margarita Pérez[b], Nuria Garatachea[e], Alejandro Lucia[b,f,*]

[a] Faculty of Sports Sciences, University of Castilla-La Mancha, 45071 Toledo, Spain
[b] European University, 28670 Madrid, Spain
[c] Sports Faculty, Pablo de Olavide University, 41013 Sevilla, Spain
[d] INEF, Universidad Politécnica de Madrid, 28040 Madrid, Spain
[e] Faculty of Health and Sport Science, University of Zaragoza, 22001 Huesca, Spain
[f] Research Institute 'i + 12' (Hospital 12 de Octubre), 28041 Madrid, Spain

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Abstract

Background: Physical activity (PA) is recommended as part of the treatment regimen for cystic fibrosis (CF) although objective methods have been scarcely used to monitor achievement of PA guidelines.
Methods: PA was measured by accelerometer in outpatient CF children (n = 47) and results were compared with those obtained in age- and gender-matched healthy controls (n = 39).
Results: 2.1% of the outpatients fulfilled PA guidelines (i.e. ≥60 min·day^{-1} of moderate-to-vigorous PA (MVPA)) vs. 34.2% of controls. Overall, lower MVPA levels were observed in CF patients than controls despite the former undergoing more light or total PA. Peak oxygen uptake was also lower in the CF group than in controls (37.5 ± 7.8 vs. 43.5 ± 7.6 ml·kg^{-1}·min^{-1}, $p = 0.002$) and was correlated with MVPA and vigorous PA in the former.
Conclusions: These findings support a need to promote PA interventions (including MVPA) targeted at improving cardiorespiratory fitness in CF children.

Keywords: Physical activity; Guidelines; Cardiorespiratory fitness

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