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## **Cystic Fibrosis**

Online lecture for physiotherapy students

DEGREE PROGRAMME IN PHYSIOTHERAPY  
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<p data-bbox="295 694 414 728"><b>Abstract</b></p> <p data-bbox="295 772 1444 952">Cystic Fibrosis is a rare and hereditary disease that affects many organs but mostly the respiratory system. Because of the recent great improvements in research and treatment, the population of Cystic Fibrosis patients are growing. The treatment of the respiratory symptoms is largely supported by the physiotherapist. This thesis is made to support the role of the physiotherapist in management of CF.</p> <p data-bbox="295 996 1444 1131">The aim of the thesis was to increase the knowledge of physiotherapy students about cystic fibrosis and about the role of the respiratory physiotherapist and objectives for patients with cystic fibrosis. The objective of this thesis was to create an online lecture for physiotherapy students.</p> <p data-bbox="295 1176 1444 1310">The thesis included a theoretical part and a practical part. It started with gathering information about Cystic Fibrosis, the pathophysiology, and the treatment associated with the disease. Then research was done about the role of the physiotherapist and guidelines were created.</p> <p data-bbox="295 1355 1444 1489">The practical part included an online class addressed to the physiotherapy students of the Satakunta University of Applied Sciences Degree Program in Physiotherapy. The online class was one hour long and included basic knowledge about Cystic Fibrosis and important knowledge about the physiotherapist's role.</p>		
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## LIST OF SYMBOLS AND TERMS

CF	Cystic Fibrosis
ACT	Airway clearance technique
ACBT	Active cycle of breathing techniques
PEP	Positive expiratory pressure
AD	Autogenic drainage
FDA	Food and drug administration
CFTR	Cystic Fibrosis Transmembrane Conductance Regulator
OPEP	Oscillating Positive expiratory pressure
MSK	Musculoskeletal

## 1 INTRODUCTION

In 1989 the gene responsible for Cystic Fibrosis (CF) disease was discovered, it is situated on chromosome 7 and is responsible of Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) dysfunction (Mazurst, 2017). The research on this disease has greatly increased the life expectancy of those suffering from it. The life expectancy went from five years in 1960 to fifty years nowadays (Bell et al., 2020; Cystic Fibrosis Foundation Web Site, 2020; Inserm Web Site, 2021). Currently, according to the Cystic Fibrosis foundation registry (2020) there are 31 411 individuals with CF.

People with cystic fibrosis have seen a vast improvement on treatment affecting their health (Bell et al., 2020). The finding of a new medicine in 2012 was a turning point for the research on this disease. Ivacaftor is approved by the food and drug administration (FDA). This is the first drug that is address to the basic causes of CF. This new drug discovery will open lot of doors to the research. (Cystic Fibrosis Foundation Web Site, 2012)

Because of the significant improvement in research and treatment, the population of CF patients is growing (Athanzio et al., 2017). The treatment of the respiratory symptoms is supported by the physiotherapist (Mazurst, 2017). This thesis is made to support the role of physiotherapist in management of CF and to increase the knowledge of physiotherapy students.

Exercising is now an important part of the management of CF and is used as medical treatment for CF in addition to the air clearance techniques (Rand et al., 2013).

## 2 AIM AND OBJECTIVES

The aim of the thesis is to increase the knowledge of physiotherapy students about cystic fibrosis and about the role of the respiratory physiotherapist and objectives for patients with cystic fibrosis. The objective of this thesis is to create an online lecture for physiotherapy students.

## 3 CYSTIC FIBROSIS

CF is a hereditary disease, autosomal recessive, which means that the child received 2 mutated gene on each chromosome 7, one from the mother and one from the father; This disease is considered as a monogenic disease because it affects only one gene. (Mazurt 2017).

There is one chance on four to have the disease for a child whose both parents have mutated gene. The two parents are carrier but not affected by the disease. If they have four children it is more likely that one will have zero affected gene, two of them will carry one affected gene (and so become carrier like their parents) and one will have the two affected genes and thus they will be affected by the CF disease (see figure 1)(Goldstein & Prystowsky, 2017).

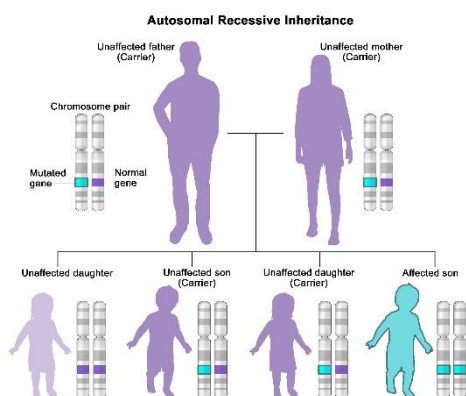


Figure 1: Autosomal recessive (National cancer institute, 2020)

### 3.1 Pathophysiology

The cystic fibrosis transmembrane conductance regulator (CFTR) is the one affected by the mutation of the gene on chromosome 7. The dysfunction of the CFTR channel can be due to an insufficient number of channels, a malfunction of the channel or even the inexistence of the channel. The CFTR protein functions as a chloride channel. In the lungs, it moves chloride ions from the interior of the cell to the exterior of the cell. Chloride is a component of salt, the chloride ion is negatively charged and will attract the positively charged side of water, so once the chloride is out of the cell it attracts water. Water helps the cilia inside the lung to move forward and backwards and hydrates the mucus on those cilia. Those cilia are moving the mucus up to remove it from the lungs. When there is a problem with CFTR protein the chloride is trapped inside the cell. So, the water, that is hydrating the mucus in the lung, is not attracted. This leads to the thickening of the mucus, that becomes dehydrated making it hard to expectorate. Mucus in our lungs is trapping the bacteria which will then be moved out of the lungs. If the mucus is too thick to be moved out, the bacteria will stay in the lungs and infect them. Deterioration of the lungs and infection in the lungs are the biggest problem for CF patient and will lead them to early death. (Cystic Fibrosis Foundation Web Site, n.d.)

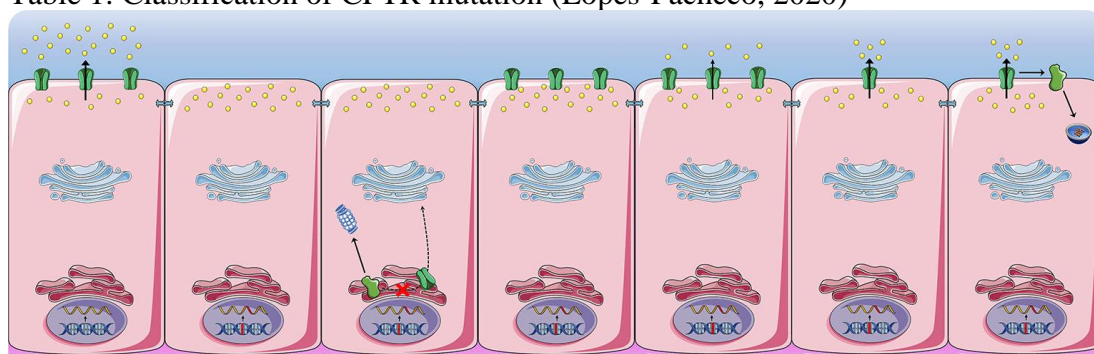
### 3.2 Different mutations

Nearly 2000 mutations have been identified (de Boeck et al., 2014), and categorised in 6 different classes. The classification is done according to the type of deficiency of the CFTR gene. In the first class the CFTR gene is not synthesized, in class II the CFTR gene is combined but not mature. In class III the CFTR gene is synthesized and transported to the plasma membrane but its regulation and/or activation is interrupted. In class IV the CFTR gene is synthesized, transported to the plasma membrane but the conductance of chloride through the channel is reduced. In class V, CFTR is synthesized but in too small a quantity. Finally, in class VI, CFTR is functional but not stable at the cell surface. (Lubamba et al., 2012) According to the

CF foundation registry (2020), the mutation F508del is the most common one and is part of the class II.

There is no information referencing a change in treatment from a physiotherapist point of view, but it is important to understand that there are various categories in the CF disease because the medical treatments and the pancreatic involvements differ (see Table 1) (Alfonsi, 2012; Pranke et al., 2019).

Table 1: Classification of CFTR mutation (Lopes-Pacheco, 2020)



	WT-CFTR	I	II	III	IV	V	VI
<i>Mutation examples</i>		G542X, R553X, R1162X, W1282X	G85E, I507del, F508del, N1303K	S549R, G551D, G1349D	R117H, R334W, D1152H	A455E, 2789+5G>A, 3849+10kbC>T	r.F508del, Q1411X
<i>Defect types</i>		Protein synthesis	Protein traffic	Channel gating	Channel conductance	Protein abundance	PM protein stability
		Minimal function				Residual function	
<i>Clinical features (global aspects)</i>		More severe phenotype				Milder phenotype	

### 3.3 Organs concerned

We can consider that all the organs affected by the disease have exocrine glands. Glands that are producing secretion that is release into a tube to reach an epithelial surface. (Freeman et al., 2022) The most common organs affected by CF are the lungs, but CF disease also affects many other organs in the body like sinuses, skin, pancreas, intestine, liver, and reproductive organs (see figure 2) (NIH, 2020).



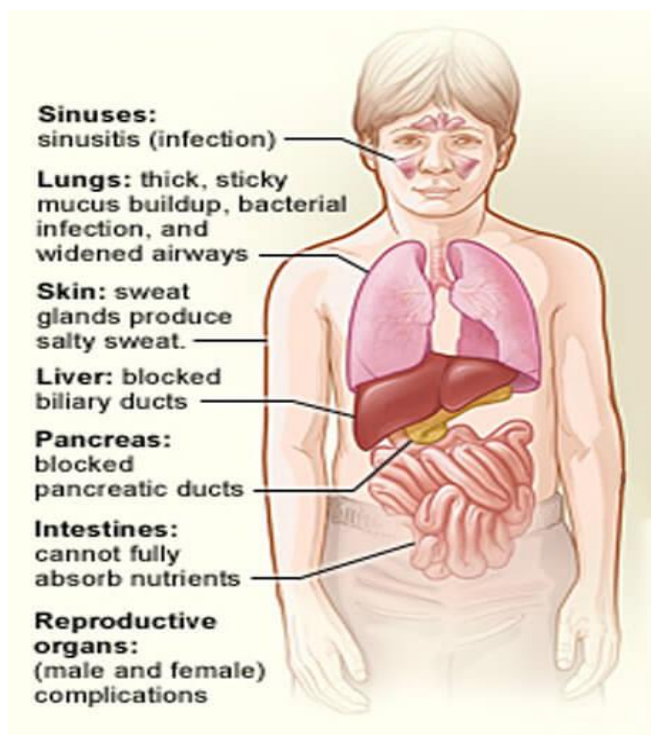


Figure 2: Organs affected by cystic fibrosis (NIH, 2020)

### 3.4 Lung infections/chronic airway infections

As explained before, the deficient CFTR gene leads to a fragile atmosphere in the lungs, a few types of bacteria or “bugs” are identified to be dangerous for the respiratory system of CF patients. *Staphylococcus aureus* (*S. aureus*) and *Pseudomonas aeruginosa* (*P. aeruginosa*) are the most common and responsible for the deaths of many patients. Lung infection is the main reason of exacerbation and leads to death, 50% of CF related deaths are due to respiratory cause (Cystic Fibrosis Foundation Web Site, 2020). These bugs can become chronic infection for many patients because they are resistant to antibiotics. (Van den Bossche et al., 2021)

It is also recommended for CF patients to not gather because of those bugs that can be transmitted to one another and that are more dangerous for CF population than healthy population. This is called a cross-infection. (Cystic Fibrosis Foundation Web Site, 2020)

The Covid pandemic has been a big worry for the Cf population but the numbers around the world show that CF patients are not more likely to get Corona Virus when compared to the general population. (Burgel & Goss, 2021)

### 3.5 Treatment

We can classify the treatment in three different classes. The respiratory treatment, the nutritional and digestive care, and the therapeutic education. Each of them contains a different kind of treatment and approach (Mazurst, 2017).

#### 3.5.1 Respiratory treatment

Cleaning the nose regularly, with sea salt solution is the first step of the respiratory treatment. The nose is used as a natural filter for the lungs, it also warms up and moistures the inhaled air. Therefore, it is important to have clean air conducted in the nose. Inhalation therapy of recombinant human deoxyribonuclease and the use of inhaled hypertonic saline (De Boeck & Amaral, 2016) are used to fluidify the mucus in the lungs. Physiotherapy, including airway clearance techniques and physical activities is also part of the treatment. We will be discussing the subject in the next chapters. It should be noted that supplemental oxygen should be readily available in case needed during physical activity or in some cases permanently. Antibiotics are used to fight the lungs infections and recently a new treatment targeting the base of CF disease is used for many CF patients. This therapy is addressed to correct the CFTR deficiency. All the vaccines, mandatory and recommended, are highly preconized for CF patients, like the influenza vaccine at age of 6 months. If the infections had affected the lungs too much, lung transplantation would be an option for the patients (Bell et al., 2020; Corvol & Clément, 2018; Mazurst, 2017).

#### 3.5.2 Nutritional and digestive care

CF disease may affect the digestive system therefore it is important for the patient to have access to digestive care. The nutrients may not always be absorbed in the system, CF patients may need some food supplements. Patients are most of the time followed by nutritionist that assesses the change of condition (Mazurst, 2017). Pancreatic enzyme supplement and micronutrients supplements may be prescribed if

the doctor notices a malabsorption of vitamins and/or minerals. (Corvol & Clément, 2018)

### 3.5.3 Therapeutic education

CF is a lifelong disease that touches many organs. The patients and their parents must understand the disease to be able to take control of it. Understanding the disease and the genetics around it is the first step. The hygiene rules need to be understood to then be able to be explain to all the people surrounding the patient. As the patient is overly sensitive to lung infections hygiene rules are for example, wearing a surgical mask, washing hands, keeping two-meter distance with other people, and avoiding contact with other CF patients. Parents and patients must know the possible complication and what to do in case of exacerbation. Patients need to have particularly good knowledge about their treatment (medicine, airway clearance, physiotherapy, nutrition) and why they have it. The awareness of the body will be a big part of the CF patient life and the patient will be the main character in the treatment of the disease. The therapeutic education gives the opportunity to share the decision making between the health professional and the patient/family (Corvol & Clément, 2018; Mazurst, 2017).

## 4 PHYSIOTHERAPY FOR CYSTIC FIBROSIS PATIENTS

Physiotherapy is a big part of CF treatment. Airway clearance and exercises have been recommended by many guidelines and will be described deeply later.

### 4.1 Objectives of the physiotherapy

As a physiotherapist it is important to keep in mind the objectives of the therapy. For CF treatment, the physiotherapist is trying to prevent and treat exacerbations, they

also have a role in treating and slowing down the deterioration of the respiratory system. The education about the disease and the treatment is an important part of the objectives. The combination of all these factors will aim to improve the lifestyle of the patient (Mazurst, 2017).

## 4.2 Physiotherapy methods

Physiotherapy for CF patients is mostly about respiratory treatment but also concerning musculoskeletal (MSK) care. The prescription of physiotherapy is made when the diagnosis is done and will slowly become a daily, or even multiple times a day treatment (Corvol & Clément, 2018).

### 4.2.1 Airway clearance technique (ACT)

Research has tried to prove what the best airway clearance is to use for patients with cystic fibrosis but there is no unambiguous evidence that one is better than the others. The choice of airway clearance is made by the practitioner and the patient. What works best for the patient, which one is their favourite, and which one is the easiest to perform (Button et al., 2016; Wilson et al., 2019). Considering this, the physiotherapist needs to know how to perform every different method of airway clearance. (Smyth et al., 2014)

Active cycle of breathing techniques (ACBT) is a method that can be used at all stage of the disease and can be introduced from the age of 2. It is an approach that can be performed individually after being taught by the physiotherapist, there is no need for materials, so it is easy to be done at home and there are little contraindications. Positive expiratory pressure (PEP) and oscillating positive expiratory pressure (OPEP) techniques envelop different methods that create resistance when exhaling, it can be a tube, a bottle with water, or a mouthpiece. Specially designed oscillation devices such as an Acapella, an Aerobika or a Flutter device may also be used. The level of resistance should be assessed regularly and changed if necessary. PEP can be used by kids and individually. The preference and

difficulty of use is taken into consideration for the choice of the device. The correct cleaning technique of the devices must be well explained and executed. Autogenic drainage (AD) is first explained by a qualified physiotherapist and can then be performed individually. It has few contraindications and can also be performed with kids, from the age of eight. Prior to the age of eight we can do an assisted AD. This technique is also beneficial for people with sensitive airways. Combining all the techniques and adapting them is possible. It is essential to not fatigue the patient and make sure they are not out of breath. The techniques can be done individually but will always be performed during the session with the physiotherapist (Morrison & Parrott, 2020). Exercises can be used as airway clearance but do not replace it. (Ward et al., 2019)

#### 4.2.2 Exercises

In 2012, two studies tried to demonstrate the importance of exercises for CF patients. This study followed children with CF for 12 months. The results showed that exercises improve the life quality of patients, but also decrease the number of exacerbation episodes so patients reported less days at hospital and an increase in the intake of antibiotics (Ledger et al., 2012; Urquhart et al., 2012).

One study in 2017 demonstrated that combined training (aerobic and resistance training) had an impact on glycaemic control for patients with CF disease, which is important because many CF patients are diagnosed with diabetes. (Beaudoin et al., 2017). Endurance training has also been proved to improve the quality of the lungs for CF patients. (Kriemler et al., 2013)

Recommendation for exercises depends on the age of the patient. However, it is recommended to have physical activity, aerobic activity, resistance training practice and specific kind of focus (like balance, coordination, control of movements, body awareness) for every age group, but with different focuses and timings (see table 2) (Castellani et al., 2018; Morrison & Parrott, 2020). Annually exercise testing is highly recommended to evaluate the cardiorespiratory performance, the lungs, and the fitness of the patient. (Morrison & Parrott, 2020). Lastly, it is not recommended

for CF patients to stay inactive; they should limit as much as possible the time they spend inactive, even during times when they feel clinically good. (Athanasio et al., 2017; Morrison & Parrott, 2020)

Table 2: Exercise training recommendation (Morrison & Parrott, 2020)

Type of activity	1-6 years	7-12 years	13-19 years	> 19 years
<b>Habitual PA</b>	60 min/day Developmentally appropriate activities	60 min/day Variety of activities enjoyed, preferably as a family	60 min/day Variety of activities enjoyed, especially with family and friends	150 min (preferably 300)/week Variety of activities of choice
<b>Aerobic exercise</b>	No formal program recommended – but should perform full-body activities that increase breathlessness and heart rate	30-60 min MVPA/day (at least 70% HR <sub>max</sub> ) Especially if using for airway clearance (must also huff/cough)	30-60 min MVPA/day (at least 70% HR <sub>max</sub> ) Especially if using for airway clearance (must also huff/cough)	30-60 min MVPA/day (at least 70% HR <sub>max</sub> ) Especially if using for airway clearance (must also huff/cough)
<b>Resistance training</b>	No formal program recommended – but should perform activities using bodyweight to develop strength (eg calisthenics)	Exercise with own body weight aimed at strengthening muscles and bones (eg calisthenics) most days. If interested, begin formal weight training under good supervision, focusing on learning good technique first (2 times / week)	Formal RT 2-3 session/week per muscle group 1-3 sets 8-12 reps 70-85% 1-RM Incorporate limb and trunk muscles	Formal RT 2-3 session/week per muscle group 1-3 sets 8-12 reps 70-85% 1-RM Incorporate limb and trunk muscles
<b>Other outcomes</b>	Encourage normal motor development, including agility, balance and coordination	Encourage normal motor development, including agility, balance and coordination	Encourage muscle activities to help prevent/minimise postural control	Adapt for disease-related complications (eg, CFRD, low bone density)

### 4.2.3 Musculoskeletal physiotherapy

The chronic respiratory diseases that are facing CF patients have an impact on the MSK system, such as tightness in the respiratory muscles or spinal kyphosis. (Lambrechts et al., 2021) Many women can face problems with pelvic floor muscles, it is also advised for CF patient to have pelvic floor rehabilitation if needed. (Corvol & Clément, 2018). Patients should have an annual MSK assessment from the age of 7 that includes posture, movement, and pain. (Morrison & Parrott, 2020)

## 5 THESIS PROCESS AND METHODS

This thesis was ordered by Satakunta University of Applied Sciences Degree Program in Physiotherapy. The material will be used as teaching material as part of the Cardiopulmonary course.

### 5.1 Schedule

Table 3: Schedule of the thesis process

Start of the thesis process	Spring 2021
The choice of a topic	October 2021
Setting objectives and aims; creation of plan; Finding literature	October - November 2021
Thesis plan presentation	11 <sup>th</sup> November 2021
Research and writing process	December 2021 – January – February 2022
Creation of course for PH20	March – April 2022
Presentation of Cystic Fibrosis course	29 <sup>th</sup> April 2022
End of writing process	May – August 2022
Final presentation	September 2022

The author has a personal interest in respiratory physiotherapy and paediatric therefore she wanted to write about those topics. Those two topics are often linked to

each other, the author has noticed that rare paediatric diseases are very often affected by respiratory symptoms. After exchanging and after confirmation with the thesis coordinator, the author noticed that cystic fibrosis was mentioned but not in depth in the cardiopulmonary and paediatric courses in the degree program of physiotherapy. A better teaching opportunity was presented when the author offered to do an oral presentation for educational support for students as the author is more fluent orally than in writing. It was a viable choice to make to do a live class. The author also feels if there is a lot of information it is easier to catch the attention of the audience in live than with text. All the permissions to do so were obtained and confirmed and the research and writing process of the thesis begun.

## 5.2 Methods

The thesis includes a theoretical and a practical part. This thesis is a practice related type of research. It exists two types of practice research: Practice based research and Practice led research. This thesis concern more the practice led research because it is considered as research aiming to obtain new knowledge to improve the practice of a subject (Candy, 2006).

Inclusion criteria was articles done from 2012 to now. The author decided so because 2012 has been a turn in CF history. The finding of a new medicine in 2012 was a turning point for the research on this disease. Ivacaftor being approved by the FDA. This is the first drug that is addressing the basic cause of CF. This new drug discovery will open lot of doors to the research. (Cystic Fibrosis Foundation Web Site, 2021)

A lot of guidelines about cystic fibrosis exist but the author has decided to take into consideration French, UK, and Australian guidelines. The author made this choice because they are part of the countries which report the most cases of CF. (Scotet et al., 2020)

Research has been done in two languages, French and English because the author is bilingual. Exclusion criteria was articles in other languages to French and English,



and research done before 2012. The data based used were Scholar and Elsevier and the search terms used were “cystic fibrosis” or “mucoviscidose;” “physiotherapy” or “kinésithérapie;” “guidelines.”

## 6 IMPLEMENTATION OF THE LECTURE

The lecture was created between January and April 2022. It was done on power point (see appendix 1) and was held via Hill. The presentation to the class was done on 29<sup>th</sup> of April. The author was doing a clinical practice in her home country at this time which is why the lecture was help via hill. The author used Padlet to ask for open anonymous feedback from the students. The Padlet was named Cystic Fibrosis feedback about the presentation (see appendix 2).

The course is one hour long, and it was possible for the students to ask questions throughout the entire lecture. The class began with a basic introduction to the disease. The author explained which gene is affected by the disease and how the disease is transmitted. The author then goes into detail about the pathophysiology, how the gene affected is affecting the lungs. The author decided to compare the CFTR channel of a healthy subject with the CFTR channel of a CF patient. Before mentioning the physiotherapy treatment, the multidisciplinary approach was mentioned, as well as the treatment of respiratory affection, digestive care, and musculoskeletal treatment. The physiotherapy part is composed of the objectives, the different type of treatment and the exercises recommendation worldwide.

At the end of the presentation, the author encouraged the physiotherapist students to open their microphones and ask questions. Many students had some questions about the condition, both relating to the physiology and the physiotherapy treatment methods, some also asked to the author for personal anecdotes about the condition. Following this the author was thanked for the presentation. Before ending the meeting, everyone was asked to take 5 minutes of their time to answer the cystic fibrosis feedback with the link sent to the chat in Hill.

Thirty people were present in the course, out of these thirty, eighteen gave feedback. The eighteen pieces of feedbacks received by the author said it was a great presentation. Of those eighteen feedbacks two asked to receive more details about life expectancy, six said the presentation was well structured, eleven mentioned the presentation was informative and eight said it had improved their knowledge on the subject.

After the author read all the feedback, the whole class received an email from her with the PowerPoint presentation attached and information about the life expectancy. The author also gave the class the opportunity to ask more questions if needed and gave thanks for their active participation and feedback.

## 7 DISCUSSION

The aim of the thesis was to increase the knowledge of physiotherapy students about cystic fibrosis, about the respiratory physiotherapist's role and objectives for patients with cystic fibrosis.

To reach this goal the online class began with a basic introduction to the disease. The author explained which gene is affected by the disease and how the disease is transmitted. The author then goes into detail about the pathophysiology, how the gene affected is affecting the lungs. The author decided to compare the CFTR channel of a healthy subject with the CFTR channel of a CF patient. Before mentioning the physiotherapy treatment, the multidisciplinary approach was mentioned, as well as the treatment of respiratory affection, digestive care, and musculoskeletal treatment.

It was important for the author to make the impact of the disease on the body clear prior to talking about the physiotherapist's role. In the opinion of the author, it helped the students to understand their objectives with the treatment. As a student herself,

she knows the problematic of focusing for the duration of the entire class, so the author also emphasised instructing with simple words and giving images and comparisons.

According to the feedbacks from the physiotherapist students, the presentation was informative, well-structured, and improved their knowledge. The author considered the class a success and her goals were met.

The author is aware that CF disease is not common in Finland, but she is also aware that physiotherapists have a noticeably significant role and impact in the treatment of this disease. It was important for her to deliver information so future physiotherapists can come back to it if one day they meet a patient in need.

The process of authoring the thesis was interesting for the author. Becoming a professional in the subject chosen is so rewarding. The complete process also gave her a better understanding about physiotherapy in general. The author became more efficient in looking for information on a subject, she also gained knowledge about the physiology of the body which helped to understand other pathologies. The confidence gained by the research and writing process and the creation and presentation of the class helped the author to feel better prepared to enter the physiotherapist world as a professional.

The author is very attracted by the education in general and it became apparent while preparing and implementing the lecture for the physiotherapy student group. The author would have preferred to do the presentation in a classroom with the students in front of her. The schedule management made it impossible. The author was in practice abroad by the time the second-year physiotherapist students were having the cardiopulmonary class. It would have been too time and money consuming to come back to the university to perform the presentation.

Respiratory physiotherapy is an intriguing part of physiotherapist work for the author, after assisting to the cardiopulmonary class it was obvious for the author to draft her thesis about this field. At the beginning of the third year of study the author had done her practice in a respiratory physiotherapy private clinic. She had been in contact with many different respiratory pathologies. She received the basic

information during class and the author understood the lack of time during studies and the limited time for every subject which prevented her from learning the topics in greater depth. In the idea to add to deepen the respiratory physiotherapy course and to pass on the knowledge learned during practice, the author decided to talk about one respiratory disease. CF was the disease that was mentioned but not explained in greater detail in the cardiopulmonary course and the author believed it deserved a better understanding and care. After choosing the subject and starting the process, CF was mentioned in the paediatric course as a rare paediatric disease that did not really need physiotherapist care. The author gave the subject of her thesis to the teacher and the teacher asked to read up on it and ensure the information be up to date. This enhances the belief that it is an important subject to write about.

The author has learned practical things during clinical practice that was not proved in literature, she found it hard to let go on this information and not talk about it in the thesis. The author found it hard to make separation with beliefs (based on experiences) that are not proved by the research. The author tried to follow the schedule of the thesis process as close as possible

For future thesis subjects, it would be interesting to read about physiotherapy with transplanted CF patients. In the experience of the author, there are specific things to have in mind to prepare the patient for the transplant, both physically and mentally as well as the patient support for after the transplant. The author did not have enough time and it was also not going towards the aim of the thesis, but she found it interesting to see that CF patients need MSK treatment, in particular the pelvic floor rehabilitation. Understanding how and what is affecting the pelvic floor muscles and what exactly is the role of the physiotherapist in that particularity for CF patient would be an interesting thesis to read for the author. Lastly, the author has found in practice, the interesting effect of the water thinning the mucus. She has looked for research on that subject but did not find anything. It may be interesting to do a trial on that subject comparing the intake of water on CF patients and the effect it has on their expectoration of mucus.

As author is planning to work in the respiratory and pediatric field in physiotherapy, she believes that the whole thesis process gave a good groundwork for her future career endeavours.

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# APPENDIX 1

### What is cystic fibrosis disease

- Rare disease
- Hereditary
- Autosomal recessive
- Management:

### Cystic Fibrosis Transmembrane conductance Regulator (CFTR) channel

- Channel channel
- Mucus chloride ions from inside to outside the cell
- Channel on a negatively charged
- Apoptosis (programmed cell death) of water

### CFTR channel in the lungs

- Channel ions are out of the cell and attract water
- Mucus becomes more liquid
- Mucus is moving the mucus to the respiratory tract
- Cells in moving out the mucus

### Deficient CFTR channel

### Classification of CFTR mutation

Class	Subclass	Frequency	Severity	Common mutation
I	Ia	~10%	Severe	ΔF508
	Ib	~1%	Severe	G542X
II	IIa	~10%	Mild to severe	R117H
	IIb	~1%	Mild to severe	S1255P
III	IIIa	~10%	Mild to severe	N1303K
	IIIb	~1%	Mild to severe	S1255P
IV	IVa	~10%	Mild to severe	R117H
	IVb	~1%	Mild to severe	S1255P
V	Va	~10%	Mild to severe	R117H
	Vb	~1%	Mild to severe	S1255P

### Organs affected

### Pathophysiological cascade of CF disease

### Lungs infection

- Pseudomonas aeruginosa
- Staphylococcus aureus
- Influenza
- Cold
- Cross infection

### Treatment

- Respiratory
- Nutritional and digestive care
- Therapeutic education

### Physiotherapy

### Prevalence of CF is growing

### Objectives

- Prevent and treat malnutrition
- Teaching and sharing about the deterioration of the respiratory system
- Education
- Improve the lifestyle of the patient

### Treatment

- Prevent and treat malnutrition
- Teaching and sharing about the deterioration of the respiratory system
- Education
- Improve the lifestyle of the patient

### Feedback

<https://forms.gle/8m8m8m8m8m8m8m8m>

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padlet

fannypoussou + 18 • 3 mois

## Cystic Fibrosis

feedback about the presentation

Very informative and great presentation. Calming voice, a lot of interesting information which you structured well and also presented really clear. Found it also really good to ask in between if there are any questions or if something is unclear. Very good job and thank you for that! Keep up the good work you are doing and all the best for your thesis! :)

**Great presentation** 🍌

Very detailed and informative presentation.

Great presentation. Easily digestible information, well researched and presented.

Nicely structured and presented. Explanations were easy to understand and improved my knowledge of the disease. Helpful to learn about treatment and physiotherapy.

Very good! Clear information, nice slides, explained very well. Thank you, Fanny!

It was really interesting and well explained presentation!

I really liked the way you explained the physiology of the disease, even though it is quite complicated. Thank you!

**great presentation !**

your presentation was really interesting. its a subject that is not well known and you made it clear to us. good structure, good speaking, it was a pleasure. thank you for that.

**Well done**

Explained with enough details and examples to help us understand. It's clear you prepared well. Thank you:)

**Interesting topic**

Very well constructed presentation about a not so much familiar disease I have learnt a lot today. I would have loved to read more about life expectancy if there's any life expectancies problem.

Your interest and motivation came through very clearly! It was a detailed presentation. Especially liked the examples from your placement and details about treatment and pt sessions. Thank you and good luck with finishing your thesis!

Thank you for lecture! Very informative and clearly spoken.

Great presentation! It helped me to understand respiratory physiotherapy and that rehabilitation can vary greatly with different conditions.

**I really liked this presentation. It was professional and interesting. You have done a huge job and have deep knowledge in this field.**

Really great job! Loved how you picked out the important information and also repeated few things. Very informative. Also I would add maybe if it's recessive or dominant and what's the life expectancy (if you did mention and I missed it, my apologies)

Interesting new knowledge for me and deep presentation many aspects

Clear and well-structured presentation. Great job!

Well structured presentation. It was well structured and the disease explained clearly. It was easy enough to grasp for someone who has no prior knowledge on the topic, but also in depth with detail of different types and their mechanisms.

The information on treatment and physiotherapy is especially valuable for us.