

Adherence to therapy in children with Cystic Fibrosis

Performance graduation research

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Block 5: Performance graduation research

Status: Concept version

Date: July 2, 2010

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Magazine: Journal of Cystic Fibrosis

Reference style: Vancouver

Number of words: 2108

Number of words samenvatting: 250

Number of words abstract: 238

Contents

1. <u>Introduction</u>	3
2. <u>Method</u>	5
2.1. Participants	5
2.2. Procedure	5
2.3. Measures	5
2.4. Statistical analyses	6
3. <u>Results</u>	7
3.1. Participants	7
3.2. Rates of adherence	7
3.3. Influencing factors	7
4. <u>Discussion</u>	9
5. <u>Conclusion</u>	11
6. <u>Recommendations</u>	12
7. <u>Reference List</u>	13
8. <u>Tables and Graphs</u>	15
9. <u>Abstract & Samenvatting</u>	18
<u>Appendix Ia</u> Questionnaire adherence	20
<u>Appendix Ib</u> Vragenlijst therapietrouw	22
<u>Appendix IIa</u> Questionnaire influencing factors	24
<u>Appendix IIb</u> Vragenlijst beïnvloedende factoren	28

1. Introduction

Cystic Fibrosis (CF) is the most common fatal hereditary disease among Caucasian people.¹ It is caused by an autosomal recessive gene that both parents must transmit and it manifests itself mainly by an abnormal secretion of the exocrine glands. This leads to sticky mucus in the airways and abnormalities in other gland products.² There still is no curative treatment for CF but through improved therapies, the survival rate increased significantly in recent years. The average age currently is 37.4 years.³ Adherence to therapy is of utmost importance for increasing the quality of life and for a successful treatment of the multiple manifestations of this disease.^{1,2,4} At a young age, parents play a major role in monitoring compliance to the multiple therapies. The relationship with the parents changes during child's development, as the child matures from being dependent towards autonomy. This is often accompanied by a decreasing adherence.^{5,6,7} Zindani et al. found that children under 12 year of age are more adherent than those older than 12 years.⁸

Adherence to therapy is defined as the extent to which the behaviour of the patient answers to the prescribed (medical and non-pharmacological) recommendations of the physician. However, the interpretation of "extent" to adherence seems to be different; 50% is seen as adherent by some researchers, while others use a cut-off of 80%.⁷ Chronically ill children show an average 50% non-adherence to the therapy which can lead to an increased morbidity and mortality.^{1,9} Adherence to therapy is influenced by many factors: behavioural, emotional, financial and psychosocial. Current literature also describes factors like: family dynamics^{2,4,9,10}, coping style, knowledge of the disease^{1,4,10}, gender^{1,2,4}, age^{1,2,4}, characteristics of therapy^{4,10,11}, socio-economic status (SES)^{1,2,9}, health condition^{4,10}, being in self-control of the therapy^{4,10}, relationship with health care providers and adverse effects of treatment¹¹. Questionnaires are most frequently used in research into factors affecting adherence. However, most are developed for specific research projects and often are not validated.^{1,11,12,13} Modi and Quittner used a list of common factors on which patients could indicate whether a mentioned factor in their opinion was important for adherence and how often it was an influencing factor.¹¹

The daily treatment of children with CF consists of several therapies: airway clearance, medication, food supplements and exercise.⁸ Research shows a difference in the degree of adherence between the different therapies.⁴ Some studies show a non-adherence to a particular therapy, while this did not apply to other therapies in the same patient.¹⁴

The measurement of adherence is often done by self-report using questionnaires or other instruments, refill information, electronic monitoring or by a telephone diary.¹⁴ None of these methods is the golden standard.⁷ A combination of these measurement is most reliable, but time consuming and expensive. The methods are sensitive to recall bias and socially desirable answers.⁴ A simple questionnaire to measure patient adherence has been developed by Myers. It specifically looks at the treatment of CF and to what extent the treatment is followed.¹²

Problem

Non-adherence is frustrating both for children and parents as well as caretakers and can lead to dissatisfaction and discouragement over the lack of a positive treatment effect. There are many factors affecting the degree of adherence in children with CF. Although many factors are described in the literature, it is unclear whether these factors also apply to children in a Dutch tertiary CF center (WKZ).

Question

To what extent are children from 6 to 12 years, who visit the outpatient Cystic Fibrosis clinic of the Wilhelmina Children's Hospital, adherent to their therapy? What factors are regarded as influencing adherence?

Purpose

This explorative study was designed to identify factors that affect adherence to treatment in children who visit the outpatient clinic of the children's hospital. This study was also meant to gain insight in the degree of adherence. Knowledge about the adherence and the influencing factors can be of great importance for both the patient and the parents.

2. Method

2.1 Participants

Participants were recruited from a tertiary Cystic Fibrosis centre, in the Netherlands. Eligibility criteria included: 1) aged between 6 and 12 years and 2) understanding and able to read Dutch. Information was sent to potential participants and their parents to give information about the study and questionnaires. Two weeks after sending the information, participants were called to remind them filling in the questionnaires.

2.2 Procedure

The design of this study was checked and approved by the Institutional Medical Ethics Committee. The participants received information about the study by postal mail. Reply to the questionnaires was seen as voluntary participation. The attached information was addressed to parents or caretakers of the child. They were asked to fill out the questionnaires together with their child.

2.3 Measures

Data were collected using the patient file and by sending two questionnaires: 1) the questionnaire “adherence” (appendix Ia) and 2) the questionnaire “influencing factors” (appendix IIa). The questionnaire “adherence” was the same as used by Myers¹², and was translated using the ‘back translation’ procedure. It is a fifteen-item self-report questionnaire to measure adherence at patients with CF. The items are scored on a Likert-scale from 1 (never) to 5 (always). The options “always” and “almost always” were defined as adherent. Total adherence scores were calculated by the number of therapies for which a patient is adherent, divided by the total number of therapies, prescribed to that patient. The researcher, following previous research of Modi¹¹, under supervision of field experts, created the questionnaire “influencing factors”. Influencing factors were selected from the literature, these were discussed by the researcher and experts. Factors that could be influenced by the healthcare team or which are common for the CF population, are included in the questionnaire. The developed questionnaire includes 32 statements that are scored on a Likert-scale from 1 (totally disagree) to 5 (totally agree). The options “totally disagree” and “disagree” were coupled as “agree” and “totally agree”.

2.4 Statistical analyses

Data were descriptive and analysed using Statistical Package for the Social Sciences (SPSS). Results were expressed as mean and range for quantitative variables. Students' t-tests were done to compare means. A p value of $<0,05$ was meant to be statistically significant.

3. Results

3.1 Participants

Data was collected in the period from January until March 2010. Sixty-seven CF patients were asked to participate in this study; four eligible participants were not included for the following reasons: (a) three patients were a-symptomatic and received no treatment at this moment, and (b) one patient was not able to read the Dutch language. Forty-two patients (66.7%) replied the questionnaires, with an age range of 6 to 12 years (mean 8.9 years) and 19 females (45.2%). Reasons for not returning the questionnaires were: lack of time, many research requests, living abroad and not receiving therapy at this moment.

3.2 Rates of adherence

Adherence rates for the different components of the CF treatment are presented in Table 1. For enzyme intake and antibiotics the adherence rate is 100 percent. No one was using insulin for diabetes and only two children used steroids. Overall rates of adherence were calculated across all treatments, it shows that the children are for 86,1% adherent to their total therapy, with a minimum of 33,3% and a maximum of 100%. There were no significant gender or age differences. The children receive an average of 9.2 therapies, with a minimum of 5 and a maximum of 15 therapies.

3.3 Influencing factors

The items of the questionnaire “influencing factors” can be divided into seven main factors: knowledge, health, parents, caregiver, adverse effects, control and time (Table 2). Some 73.8% of the children have the opinion that they will be more adherent with increasing knowledge about the disease; this is 69.0% when regarding the therapies. Most patients feel healthy at the moment of this study (78.6%). When feeling ill, 83.3% take their medicine and 52.4% do their exercises. When patients feel healthy, they more often forget their treatment (21.4%); 26.1% is neutral in this statement. About 88.1% of the parents prepare the medication for their children and 76.1% stimulate their child to do the exercises. Percentages of parents controlling their child for taking the medication is 88.1%, for exercises 71.4%, and diet appointments 64.3%. Three children (7.1%) dislike parents intervening with the complete treatment; 21.4% is neutral in this statement. The relationship with the doctor is good in 100%, for physiotherapist it's 81.0%, and for the dietician 78.6%. Some 61.9% of the children experience the side effects from the medication as annoying. Children have little control over their treatment: taking the medication themselves is 31.0%; 92.9% does not determine themselves which medication to take at which time; for doing the exercises this is 69.0%.

Taking medication and doing exercises takes a lot of time says 33.3% of the children. Fifty percent of the children take time to complete the treatment, 42.9% are neutral in this statement. From all children, 66.7% can do exercise or daily sports as recommended.

4. Discussion

Main purpose of this pilot was to identify which factors influence adherence at children with CF, and to measure the adherence rates. The results of this pilot study can be used to design a validation study into the Myers' questionnaire.

Results

Previous research shows low response rates (39.8%)¹⁵. In our research the response rate is quite high. This could be caused by the fact that questionnaires were filled in by the parents and the researcher phoned the participants several times to complete the questionnaires. Reasons for not returning the questionnaires were: lack of time, many research requests, living abroad and child receiving no therapy.

The adherence rates per treatment seems to differ, the lowest rate is 31,6% for overnight feeding and the highest 100% for enzyme intake and antibiotics. The level of total adherence is high (86,1%), with a minimum of 33,3% and a maximum of 100%, compared with other studies, which reported levels of 30-70%¹.

Parents seem to be important in taking and preparing the medication, and they do control their children in taking the medication and doing the exercises. Children ask little support in their treatment, most support is asked from parents; less from the doctor, physiotherapist and dietician.

Limitations and strengths of the study

The population of this study consists of children aged between 6 and 12 years. They are not fully capable to complete the questionnaires themselves, so this is done with support of their parents. This could have led to desirable answers. Interestingly, most missing values were found in questions and answers about the parents.

Social desirability plays a major role in measuring adherence. Patients want to satisfy the expected behaviour, even if they are non-adherent to the treatment. Self-reported measures of adherence tend to be overestimated; adherence rates would probably be lower¹².

The questionnaire developed and used by Myers was used to measure adherence. It is known that adherence rates are overestimated when self-reported. To increase the validity, other methods can be used to determine the adherence, such as refill information, telephone diaries or electronic monitoring.¹⁴ Because these methods are time consuming and expensive to perform, we have chosen for the simplest method to measure adherence.

Reliability and validity

To determine which factors are influencing adherence to treatment, a specific questionnaire must be developed. Modi and Quittner formulated the most common influencing factors; patients can choose any factors that were relevant. Next, participants rated the frequency in which these factors occurred. In this study factors were selected from the literature, and statements were created to measure how patients experience influencing factors.

No previous study has used the questionnaire “influencing factors” used in this study. The researchers, based on a literature study, have developed this questionnaire specifically for this pilot- study. The results can be used in other CF centres, although it must be mentioned that the questionnaire has yet to be validated.

5. Conclusion

Children in this study seem to be very adherent (86,1%), but for specific treatments the rates are lower. For enzyme intake with meals and antibiotics intake, the adherence rates were 100%. The lowest adherence is for overnight feeding (31.6%).

In each influencing factors there is a large range in response, all categories were scored. Parents seem to be important in taking and preparing the medication, and parents do control their child in taking the medication and doing the exercises. Children ask little support in the treatment. Most support comes from the parents, less from doctor, physiotherapist and dietician.

6. Recommendations

Health care providers who treat children with CF should take into account that every child experiences other factors as influencing the adherence. Therefore the treatment of a child should be focussed on this specific child.

When the questionnaire is validated, a study can be done to increase adherence in children with CF.

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8. Tables and Graphs

Table 1: Percentage of adherence per treatment.

Treatment	Part of treatment N	Adherent N (%)
Enzymes (meal)	40	40 (100)
Enzymes (snacks)	40	36 (90)
Vitamin supplements	40	38 (95)
Exercise	42	40 (95.2)
Cancelling outpatient appointments	42	41 (97.6)
Overnight feeding	19	6 (31.6)
Antibiotics	39	39 (100)
Bronchodilators	19	16 (84.2)
High-fat diet	35	26 (74.3)
Physiotherapy	28	19 (67.9)
Steroids	2	1 (50.0)
Regular supplements	9	4 (40.0)
Insulin	0	0 (0)
DNase (Pulmozyme)	21	18 (85.7)
Thin bone medication	7	5 (71.5)
Total treatment		86,1

Table 2: Influencing factors

Factor	Disagree	Not agree / Not disagree	Agree
Knowledge			
- I have much knowledge about my disease.	11,9	31,0	57,1
- If my knowledge about the disease increases, I understand better why I need to follow the therapy.	7,1	19,0	73,8
- I have much knowledge about my treatment.	7,1	38,1	54,8
- If my knowledge of the treatment increases and I understand why something is needed, I will perform better.	9,5	21,4	69,0
- I know what medication I should use.	2,4	14,3	83,3
- I know wherefore the medication I use is.	0,0	35,7	64,3
Health			
- At this moment I feel very healthy.	4,8	16,7	78,6
- When I feel good I forget my treatment.	52,4	26,2	21,4
- When I am sick, I take my medicine.	11,9	4,8	83,3
- When I am sick, I do my exercise.	18,4	23,7	57,9
Parents			
- My parents prepare my medication.	2,4	9,5	88,1
- My parents encourage me to do my exercises.	8,1	5,4	86,5
- I ask my parents to support me with my treatment.	10,0	25,0	65,0
- My parents check my medication occupy.	2,4	9,5	88,1
- My parents sure I do my exercises.	5,3	15,8	78,9
- My parents sure I keep my diet appointments.	15,4	15,4	69,2
- I dislike when my parents interfere with my treatment.	69,2	23,1	7,7
Caregiver			
- I ask my doctor to aid in my treatment.	28,2	41,0	30,8
- I ask my physiotherapist to aid in my treatment.	25,0	41,7	33,3
- I ask my dietitian to aid for the nutritional support.	23,7	42,1	34,2

- My relationship with the doctor is good.	0,0	0,0	100,0
- My relationship with the physiotherapist is good.	0,0	8,1	91,9
- My relationship with the dietician is good.	2,6	12,8	84,6
Adverse effects			
- My medications have unpleasant side effects.	63,4	29,3	7,3
- Taking my medication takes a lot of time.	47,6	19,0	33,3
- Doing my exercises takes a lot of time.	28,9	34,2	36,8
- I find it difficult that I have CF.	4,8	31,0	50,0
Control			
- I concern myself that I occupy the right medication.	21,4	47,6	31,0
- I can decide myself which medication I take at which time.	92,9	2,4	4,8
- I can decide myself if I do my exercises.	76,3	18,4	5,3
Time			
- I take time for my treatment.	2,5	45,0	52,5
- I can sport or exercise daily as recommended.	7,1	26,2	66,7

9. Abstract & Samenvatting

9.1 Abstract

Background Cystic Fibrosis (CF) is the most common fatal hereditary disease among Caucasian people. There is no curative treatment for CF but through improved therapies, the survival rate increased significantly in recent years. One of the factors that play a role is adherence. Adherence is defined as the extent to which the behavior of the patient answers to the prescribed (medical and non-pharmacological) recommendations of the physician. Adherence to therapy is influenced by many factors: behavioral, emotional, financial and family.

Aim and research question The aim of this study is to identify the factors that affect adherence to treatment in children with CF. Therefore the following research question was formulated: To what extent are children from 6 to 12 years, who visit the outpatient Cystic Fibrosis clinic of the Wilhelmina Children's Hospital, adherent to their therapy? What factors are regarded as influencing adherence?

Method To 67 patients with CF, aged between 6 and 12 years, questionnaires ('adherence' and 'influencing factors') were sent. Reply to the questionnaires was seen as voluntary participation.

Results Total adherence rates were 86,1%. Parents seem to play an important role in preparing the medication as in control the intake and in supporting to do the exercises.

Conclusion Children who visit the outpatient clinic are very adherent, although it can be increased.

Recommendations Validation of the questionnaire influencing factors can be done to create a practicable instrument.

Keywords Cystic Fibrosis (CF), adherence, influencing factors, children

9.2 Samenvatting

Achtergrond Cystic Fibrosis (CF) is de meest voorkomende dodelijke erfelijke ziekte onder caucasische mensen. Er is geen genezende behandeling voor CF, maar door betere therapieën zijn de overlevingskansen de laatste jaren toegenomen. Een belangrijke factor hiervoor is therapietrouw. Therapietrouw is de mate waarin het gedrag van de patiënt overeenkomt met de voorgeschreven (medicamenteuze en niet-medicamenteuze) aanbevelingen van de arts. Vele factoren spelen een rol in de therapietrouw; gedrags-, emotionele-, financiële- en familiefactoren zijn van belang.

Doel en onderzoeksvraag In het Wilhelmina Kinderziekenhuis (WKZ) te Utrecht is niet bekend welke factoren van invloed zijn op de therapietrouw. De kinderen leven in een andere cultuur en onder een ander pedagogisch klimaat dan kinderen uit eerdere onderzoeken.

In welke mate zijn kinderen van 6 tot 12 jaar, die de Cystic Fibrosis poli van het Wilhelmina Kinderziekenhuis bezoeken, therapietrouw? Welke factoren worden aangemerkt als zijnde beïnvloedend op de therapietrouw?

Methode Aan 67 patiënten met CF tussen de 6 en 12 jaar zijn vragenlijsten 'therapietrouw' en 'beïnvloedende factoren' verstuurd. Terugsturen van de vragenlijsten is gezien als vrijwillige deelname.

Resultaten De kinderen zijn voor 86,1% therapietrouw. Ouders blijken een belangrijke rol te spelen in zowel het klaarleggen van de medicijnen als de controle op inname en de stimulans om oefeningen te doen.

Conclusie De kinderen in het WKZ zijn in grote mate therapietrouw, er zijn echter nog punten waarop verbetering geboekt kan worden.

Aanbevelingen Validatie van de vragenlijst beïnvloedende factoren is mogelijk om een bruikbaar instrument te verkrijgen.

Trefwoorden Cystic Fibrosis (CF), therapietrouw, beïnvloedende factoren, kinderen

Appendix Ia Questionnaire adherence

Questionnaire 1: Adherence

Please circle the answer that best describes you. If you are not prescribed a specific treatment please circle X.

I take my enzymes with meals.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

I take my enzymes with snacks.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

Taking vitamin supplements is part of my daily routine.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

Regular exercise is part of my life-style.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

I cancel or reschedule my outpatient appointments.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

Overnight feeding is part of my routine.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

I take antibiotics as prescribed by my doctor.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

I use my bronchodilators as prescribed.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

A high fat diet is part of my lifestyle.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

Physiotherapy is part of my daily routine.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

I take steroids as prescribed by my doctor.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

Regular dietary supplements are part of my everyday life.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

I take insulin (for diabetes) as prescribed by my doctor.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

I use DNase (Pulmozyme) as directed.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

I take medication for thin bones as prescribed.

never	hardly ever	sometimes	almost always	always	Not part of my treatment
1	2	3	4	5	X

Appendix Ib Vragenlijst therapietrouw**Vragenlijst 1: Therapietrouw**

Veel mensen die wij gesproken hebben zeggen dat ze letten op hun CF op een manier die bij hen past, in plaats van het tot op detail volgen van de aanbevelingen van behandelaars. Wij zijn benieuwd hoe jij op je CF let en hoe jij kijkt naar de verschillende behandelingen.

Geef aan in hoeverre de stellingen voor jou van toepassing zijn. Wanneer het geen deel van je behandeling is omcirkel je de X.

Ik neem mijn enzymen bij de maaltijd

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Ik neem mijn enzymen met tussendoortjes

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Het innemen van vitamine supplementen is deel van mijn dagelijkse routine

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Regelmatige beweging en sport is onderdeel van mijn leefstijl

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Ik annuleer of verplaats mijn poliklinische afspraken

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Voeding tijdens de nacht is een onderdeel van mijn routine

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Ik neem de antibiotica zoals voorgeschreven door mijn arts

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Ik gebruik mijn brochoverwijders (luchtwegverwijders) zoals voorgeschreven

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Een hoog-vet dieet is een deel van mijn leefstijl

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Fysiotherapie is een deel van mijn dagelijkse routine

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Ik neem de steroïden zoals voor geschreven door mijn arts

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Reguliere supplementen zijn deel van mijn dagelijks leven

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Ik neem insuline (voor diabetes) zoals voorgeschreven door mijn arts

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Ik gebruik DNase (pulmozyne) zoals voorgeschreven

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Ik neem de medicatie ter versteviging van mijn botten zoals voorgeschreven

nooit	bijna nooit	soms	bijna altijd	altijd	geen deel van mijn behandeling
1	2	3	4	5	X

Dank je wel voor het invullen van deze lijst, we vragen je de tweede vragenlijst ook in te vullen.

Appendix IIa Questionnaire influencing factors

Questionnaire 2: Influencing factors

Indicate the extent in which you agree with the following statements.

I have much knowledge about my disease.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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If my knowledge about the disease increases, I understand better why I need to follow the therapy.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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I have much knowledge about my treatment.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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If my knowledge of the treatment increases and I understand why something is needed, I will perform better.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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I know what medication I should use.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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I know wherefore the medication I use is.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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I concern myself that I occupy the right medication.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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At this moment I feel very healthy.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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When I feel good I forget my treatment.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

When I am sick, I take my medicine.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

When I am sick, I do my exercise.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

I take time for my treatment.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

I can sport or exercise daily as recommended.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

My parents prepare my medication.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

My parents encourage me to do my exercises.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

I ask my parents to support me with my treatment.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

My parents check my medication occupy.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

My parents sure I do my exercises.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

My parents sure I keep my diet appointments.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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I dislike when my parents interfere with my treatment.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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I ask my doctor to aid in my treatment.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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I ask my physiotherapist to aid in my treatment.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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I ask my dietitian to aid for the nutritional support.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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My relationship with the doctor is good.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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My relationship with the physiotherapist is good.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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My relation with the dietitian is good.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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My medications have unpleasant side effects.

totally disagree 1	disagree 2	not agree not disagree 3	agree 4	totally agree 5
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Taking my medication takes a lot of time.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

Doing my exercises takes a lot of time.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

I find it difficult that I have CF.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

I can decide myself which medication I take at which time.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

I can decide myself if I do my exercises.

totally disagree	disagree	not agree not disagree	agree	totally agree
1	2	3	4	5

If all is well you have now completed two questionnaires. We want to thank you for your participation.

Appendix IIb Vragenlijst beïnvloedende factoren**Vragenlijst 2: Beïnvloedende factoren**

Geef aan in hoeverre je het eens bent met de onderstaande stellingen.

Ik heb veel kennis over mijn ziekte.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Als mijn kennis over de ziekte toeneemt, begrijp ik beter waarom ik de therapie moet volgen.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik heb veel kennis over mijn behandeling.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Als mijn kennis over de behandeling toeneemt en ik goed begrijp waarom iets nodig is zal ik het beter uitvoeren.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik weet welke medicijnen ik moet gebruiken.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik weet waarvoor de medicijnen zijn die ik moet gebruiken.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik zorg er zelf voor dat ik de juiste medicatie inneem.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Op dit moment voel ik me erg gezond.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Als ik me goed voel vergeet ik mijn behandeling.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Als ik ziek ben neem ik mijn medicijnen.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Als ik ziek ben doe ik mijn oefeningen.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik neem de tijd voor mijn behandeling.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik kan dagelijks sporten of bewegen zoals voorgeschreven.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn medicijnen worden klaargelegd door mijn ouders.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn ouders stimuleren mij om mijn oefeningen te doen.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn ouders vraag ik om steun in mijn behandeling.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn ouders controleren of ik mijn medicatie inneem.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn ouders controleren of ik mijn oefeningen doe.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn ouders controleren of ik mij aan mijn dieetafspraken houd.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik vind het vervelend als mijn ouders zich met mijn behandeling bemoeien.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik vraag mijn dokter om steun in mijn behandeling.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik vraag mijn fysiotherapeut om steun in mijn behandeling.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik vraag mijn diëtist om ondersteuning bij het voedingsadvies.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn relatie met de dokter is goed.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn relatie met de fysiotherapeut is goed.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn relatie met de diëtiste is goed.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Mijn medicijnen hebben vervelende bijwerkingen.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Het nemen van mijn medicatie kost veel tijd.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Het doen van mijn oefeningen kost veel tijd.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik vind het moeilijk dat ik CF heb.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik mag zelf bepalen welke medicijnen ik in wil nemen op welk moment.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Ik bepaal zelf of ik mijn oefeningen uitvoer.

helemaal oneens 1	oneens 2	niet eens niet oneens 3	eens 4	helemaal eens 5
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Als het goed is heb je nu de twee vragenlijsten ingevuld. Wij willen je hartelijk bedanken voor je deelname!