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Telerehabilitation and telemonitoring interventions programs used to improving quality of life in people with cystic fibrosis: A systematic review

Lucía Ortiz Ortigosa[1], María Jesús Vinolo-Gil[2], José-Manuel Pastora Bernal[1], María Jesús Casuso-Holgado[3], Manuel Rodriguez-Huguet[2] and Rocío Martín-Valero[1]

Abstract

Background: Cystic fibrosis causes mucus to build up in the lungs, digestive tract, and other areas. It is the most common chronic lung disease in children and young adults. It requires daily medical care. Before the COVID-19 pandemic, telerehabilitation and telehealth were used, but it was after this that there was a boom in these types of assistance in order to continue caring for cystic fibrosis patients.

Objective: The objective is to evaluate the effect of telemedicine programs in people with cystic fibrosis.

Methods: For the search, the PubMed, Scopus, Web of Science, PEDro, Cochrane, and CINAHL databases were used. Randomized controlled trials, pilot studies, and clinical trials have been included. The exclusion criteria have considered that the population did not have another active disease or that telemedicine was not used as the main intervention. This study follows the PRISMA statement and has been registered in the PROSPERO database (CRD42021257647).

Results: A total of 11 articles have been included in the systematic review. No improvements have been found in quality of life, forced expiratory volume, and forced vital capacity. Good results have been found in increasing physical activity and early detection of exacerbations. Adherence and satisfaction are very positive and promising.

Conclusions: Despite not obtaining significant improvements in some of the variables, it should be noted that the adherence and satisfaction of both patients and workers reinforce the use of this type of care. Future studies are recommended in which to continue investigating this topic.

Keywords

Telemedicine, cystic fibrosis, telerehabilitation, exercise, telemonitoring

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Introduction

Cystic fibrosis (CF) is characterized by poor airway hydration and infection that promotes the accumulation of secretions and subsequent lung damage.[1,2] It usually includes bronchiectasis and even progresses to respiratory failure.[3] People who suffer from this disease present symptoms daily, such as follows: wheezing, fever, cough, sputum production, breathing difficulties, and pressure in the chest.[4]

[1]Department of Physiotherapy, Faculty of Health Science, University of Málaga, CTS-1071 Research Group, Málaga, Spain
[2]Department of Nursing and Physiotherapy, University of Cadiz, Cádiz, Spain
[3]Department of Physiotherapy, Faculty of Nursing, Physiotherapy and Podiatry, University of Seville, Sevilla, Spain

Corresponding author:
Rocío Martín-Valero, Department of Physiotherapy, Faculty of Health Science, University of Málaga, CTS-1071 Research Group, Arquitecto Francisco Peñalosa, 3, 29071 Málaga, Spain.
Email: rovalemas@uma.es

Patients with cystic fibrosis have numerous admissions and visits to the hospital to treat possible respiratory exacerbations that are taking place.[3] These people are often accompanied by poor quality and life expectancy, poor physical condition, depression, and anxiety.[3] Patients with cystic fibrosis need assistance from a multidisciplinary team for life in order to carry out good control of the person in all areas.[5]

Prior to the COVID-19 pandemic, telerehabilitation and telehealth were already used in people with cystic fibrosis, although it is true that due to the pandemic that emerged in 2020 and with it the need to find solutions to continue caring for patients with cystic fibrosis, there was increased use of telemedicine and telerehabilitation. Above all, these types of rehabilitation continue to be used and evaluated because they are largely beneficial to the patients themselves.[5–7]

From the Pediatric CF Service of the Royal Brompton Hospital, the multidisciplinary team and other studies report that, with the same ease, telehealth offers the same collection of information as usual care, checkups are carried out more frequently, improves access to a more specialized care.

The assistance is carried out in a more optimal way, since culture samples could be carried out prior to revisions in order to carry out a more effective treatment, revisions could be carried out more easily and with less delay after a new treatment. In addition, the cost of the patients themselves is reduced since trips to the different consultations are eliminated and the exposure to infection in hospitals of high-risk patients is eliminated.[5,6,8]

In addition, telerehabilitation offers new possibilities in different areas, for example, in physiotherapy, the possibility of reviewing the airway cleaning equipment is created and with it, advice on its cleaning (instruments that are not normally carried out for revisions). The demographic area to which a good exercise program can be facilitated can be expanded, since the necessary economic expense in face-to-face training is reduced (gyms, physiotherapists, etc.).[6]

Thanks to the great progress, evolution and the health situation that we have experienced in recent years, information technology has promoted telemedicine research. This evolution of telemedicine favors the interaction between the patient and the health system.[9] Telemedicine seeks satisfactory results with respect to the health status of patients, who, focusing on this study, suffer from cystic fibrosis and the costs associated with hospital care.[4] They are capable of creating new ways to encourage and support physical activity and detect pulmonary exacerbations early.[3,4]

Objectives

Among the objectives of the study, the main and most frequently repeated one is quality of life. Among the secondary objectives is the forced expiratory volume (FEV1). Other secondary objectives that have been assessed are 6 MINUTE WALK TEST (6 mwt), exercise capacity (peak VO2), and forced vital capacity (FVC).

Methods

This systematic review has been carried out following the recommendations of PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analysis).[6]

In addition, it has been registered in the Prospective International Registry of Systematic Review (PROSPERO).

Search strategy

The databases used for the search were PubMed, Scopus, Web of Science, PEDro, Cochrane, and CINAHL. The OR and AND booleans were used. The search strategy was carried out following the PICO model (Population, Intervention, Comparison and Result). The search strategy combines terms included in MeSH related to the population "Cystic Fibrosis" AND ("Telemedicine" OR "Telerehabilitation" OR "Computer Methodologies" OR "Telemonitoring"). Abstracts and articles were screened for further eligibility. The search strategy has followed the registry of the protocol published in PROSPERO with number CRD42021257647.

The search strategy process for the articles selected for this review can be found in Appendix 1.

Eligibility criteria

Randomized controlled trials, pilot studies, and clinical trials and articles written in Spanish or English have been included.

The exclusion criteria considered that the participants in the published studies had no other active or severe lung disease and that they did not use telemedicine as their main intervention.

Finally, 11 studies have been included, with a total of 1035 participants, whose ages range from 0 to 47 years.

Quality assessment

The evaluation of the methodological quality of the studies selected in this systematic review and meta-analysis was carried out using the Physiotherapy Evidence Database (PEDro) scale. This scale is made up of 11 criteria that assess the internal validity of the articles.[7,8]

Risk of bias of included studies

The risk of bias was calculated for each study selected using the Cochrane Collaboration Tool.[10] The following types of bias were assessed as follows: selection bias, performance

bias, detection bias, attrition bias, reporting bias, and other bias. Two reviewers (LOO and RMV) assessed the methodological quality and the risk of bias of the studies. In case of doubt, authors resolved disagreements by consensus and consulting a third author (MJVG) when necessary.

Statistical analysis

A data extraction has been carried out, which will consist of summarizing the information for qualitative synthesis. The characteristics of each of the articles have been extracted together with the interventions carried out, the variables examined, and the results. All these data have been collected in a descriptive table for the subsequent narrative synthesis.

Results

Study selection

A total of 721 articles were found. After applying the filters, we were left with 147 studies. Finally, a total of 11 articles have been included in the systematic review. The process can be seen in Figure 1.

Methodological quality of included studies

Table 1 shows the rating obtained from the articles selected after the methodological quality review, for which the PEDro scale was used. According to this scale, one of the 11 articles reached a 9, which indicates excellent methodological quality, two reviewed articles reached a score of 7,

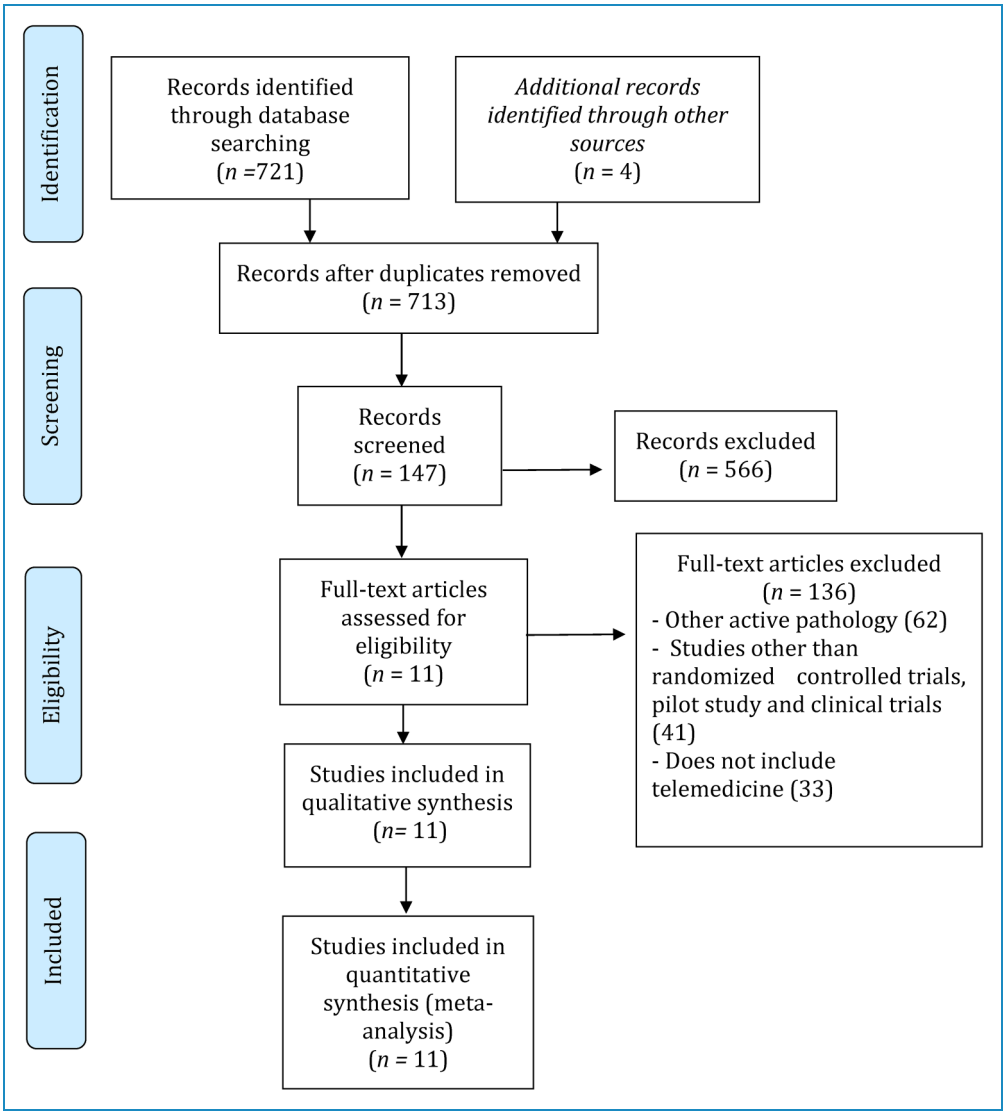

Figure 1. Search strategy.[22] For more information, visit www.prisma-statement.org.

Table 1. Methodological quality according to the PEDro scale.

Criteria	1	2	3	4	5	6	7	8	9	10	11	Total
Chrysochoou et al.[11]	N	N	N	Y	N	N	N	Y	Y	Y	Y	5
Cummings et al.[12]	Y	Y	Y	Y	Y	N	Y	Y	Y	Y	Y	9
Wilkinson et al.[13]	N	Y	Y	Y	N	N	N	N	N	Y	Y	5
Murgia et al.[14]	N	N	N	Y	N	N	N	Y	Y	Y	Y	5
Hommerding et al.[15]	N	Y	N	N	N	N	N	Y	Y	Y	Y	5
Grzincich et al.[16]	Y	Y	N	N	N	N	N	Y	Y	N	N	3
Rovedder et al.[17]	Y	Y	Y	Y	N	N	N	Y	Y	Y	Y	7
Bathgate et al.[18]	Y	Y	N	Y	N	N	N	Y	Y	Y	Y	6
Hork et al.[19]	Y	N	N	Y	N	N	N	Y	Y	Y	Y	5
Gur et al.[20]	N	N	N	Y	N	N	N	Y	Y	Y	Y	5
Wood et al.[21]	Y	Y	N	Y	Y	N	N	Y	Y	Y	Y	7

PEDro: Physiotherapy Evidence Database. The eligibility criteria do not contribute to the total score. Y: yes; N: no.

which is considered evidence level 1, one obtained a score of 6, and six studies obtained a score of 5, which is considered level of evidence 2. Finally, one study obtained a score of less than 4, which indicates poor methodological quality.

Risk of bias of included studies

The Cochrane Risk of Bias Assessment Tool was used to assess the risk of bias of the articles included in this review. The results of the risk of bias can be seen in Figure 2.

It should be noted that with respect to "selective reporting bias" and "other bias," all articles were of unclear risk (Figure 3). Regarding "selective information bias," only one of the articles had published the protocol retrospectively, so the variables to be measured could not be known in advance. As for "other risks," biases could appear due to the type of study, almost all of which are observational, with small samples, without randomization. Certain biases could also appear because access to the Internet is more accessible or easier for some patients."

Study design

Regarding the main characteristics of the different articles (Table 2), it can be seen that a total of 664 subjects were evaluated by the selected studies. The article that used the largest sample size was Chrysochoou et al.,[11] while those who used the smallest sample size were Wilkinson et al.[13] and Murgia et al.[14] The ages of the participants were between 5 and 47 years old, with three articles in which it was not specified.[10,11,14,17,21,22] The longest intervention duration was 4.5 years,[14] and the shortest was 2 months.[18] Two of the articles do not indicate the duration of the intervention.[11,16] Regarding the type of intervention used, as well as the variables that each article studies, can be seen in Table 2.

Quality of life

The questionnaires used to assess quality of life have been the CFQ—Cystic Fibrosis Questionnaire[13,15,17,18,20] and the SF-36 Questionnaire.[17]. In a protocol found, another series of questionnaires is used, such as the EQ-5D-5L or EQ-5D-Y-5L in adolescents.[23] One of the articles does not indicate which questionnaire they have used.[11] However, after obtaining the results, it is shown that in most cases, there have been no significant differences in the data obtained before starting the study and after it.[13,15,17,20] Quality of life improved significantly in only two, in one with an effect size of 43.44 (14.50) ($p < 0.001$),[11] and in the other study social functioning (Cohen's $d = 0.97$) and vitality (Cohen's $d = 0.82$) had large treatment effects immediately after the intervention.[14,18]

Forced expiratory volume (FEV1)

The great majority of articles have studied it.[11,14,15,19] The results show significant improvements in one of the four

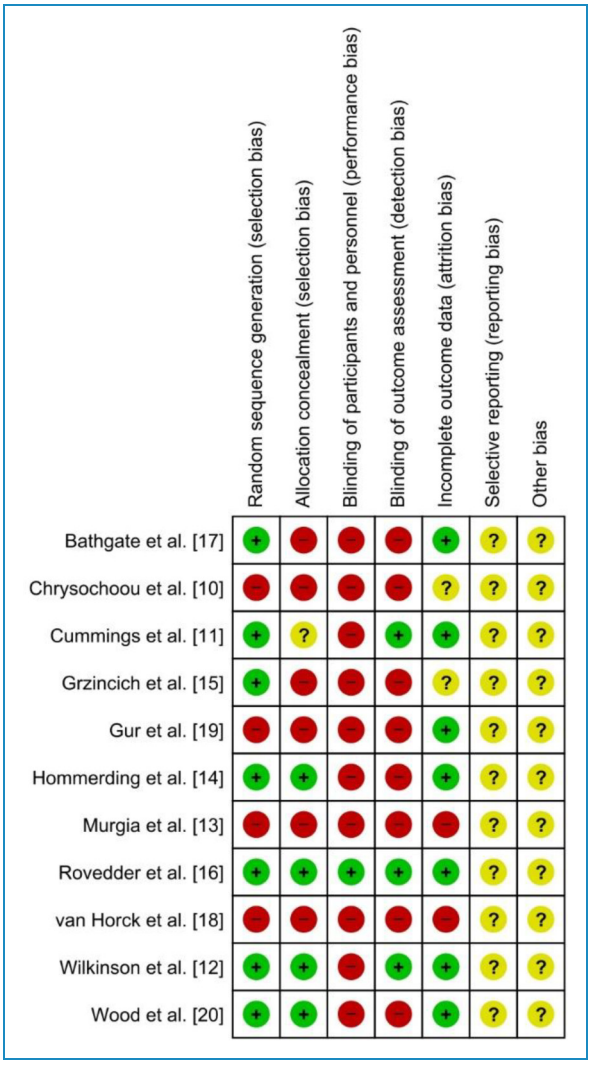

Figure 2. Risk of bias summary.

articles that analyze this value ($p = 0.0021$).[14] Two of the articles did not obtain significant improvement,[11,15] and the last one examined the FEV1 value, but comparing it between a group that presented exacerbations and one that did not. The FEV1% pred at the onset of the exacerbation was significantly lower than the FEV1% pred in the group without exacerbation (mean difference 16.3%, $p = 0.012$).[19] The combination of FEV1% pred and Respiratory Symptom Score (RSS) had a sensitivity to predict an exacerbation of 92.9% and a specificity of 88.9%.[19]

6 MINUTE WALK TEST (6 mwt), exercise capacity (peak VO2), and forced vital capacity (FVC)

Only one article of those selected evaluated the aforementioned objectives. The one who valued the 6 MINUTE WALK TEST (6 mwt) did not obtain significant differences. However, this same article also used the muscle strength test (RM) where the experimental group presented a significant improvement after 3 months in muscle strength in the 1RM test ($p = 0.011$ upper left member) and ($p = 0.029$ upper right member).[17] The article that evaluated exercise capacity showed a significant increase in the practice of physical exercise in favor of the intervention group ($p = 0.01$), but no significant improvement was observed with respect to exercise capacity (peak VO2).[15] In addition, this same article also studied the FVC, but did not obtain significant improvements between both groups[15]; regarding the increase in physical activity, if they achieved the effect size (effect size) 11 (64.7) $p = 0.01$.[11,15]

Discussion

This systematic review supports that telemedicine program could be considered at least as effective as conventional

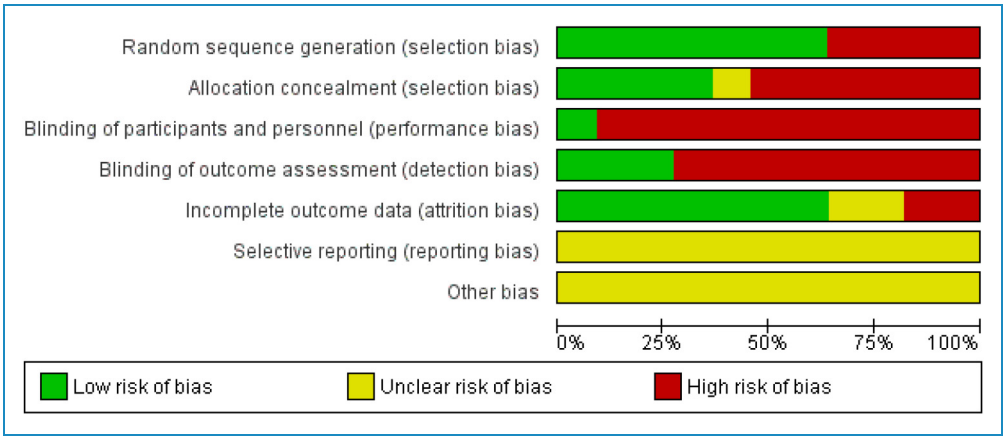

Figure 3. Risk of bias graph.

Table 2. PICO table.

Author and year	Study title	Type of study	Intervention	Variables	Results
Chrysochoou et al. (2017)[11]	Telephone monitoring and home visits significantly improved the quality of life, treatment adherence and lung function in children with cystic fibrosis.	Systematic, multicenter review. *N* = 320. Participant age: not specified. Intervention time: It does not indicate.	IG: telephone communication with health personnel. CG: Home visits by health personnel.	FEV1 (forced expiratory volume): program effectiveness.	No significant differences between the two groups in FEV1 and the days and cost of hospitalization after home care program.
Cummings et al. (2011)[12]	Enhancing self-efficacy for self-management in people with cystic fibrosis.	Randomized controlled trial. *N* = 19. Participant age: 14–47 years. Intervention time: 6 months.	G1: access to a self-efficacy program. G2: Access to the same self-efficacy program plus the provision of a mobile phone and a web application to monitor symptoms. CG: normal attention.	Subjective health status. Stanford self-efficacy for chronic disease management. Forced expiratory volume (FEV1) and forced vital capacity (FVC).	The results show challenges in stimulating self-management behaviors, particularly in adolescents, and in evaluating the role of mobile applications in their support.
Wilkinson et al. (2008)[13]	A feasibility study of home telemedicine for patients with cystic fibrosis awaiting transplantation.	Prospective pilot study. *N* = 16. Participant age: 21–38 years. Intervention time: 6 months.	IG: telemedicine (ISDN line installed at home and videoconferencing unit connected to home television). CG: standard care.	Cystic Fibrosis Quality of Life Questionnaire. Beck Anxiety Inventory. Beck Depression Inventory. Borg Anxiety Scale.	No significant differences in quality of life, anxiety levels, depression levels, hospital admissions, or clinic visits. Significant improvement in the perception of body image in favor of the telemedicine group.
Murgia et al. (2015)[14]	Telemedicine home program in patients with cystic fibrosis: results after 10 years.	Clinical trial. *N* = 16. Participant age: not specified. Intervention time: 4.5 years.	IG: telehomecare (Spirotel instrumentation) along with standard therapy. CG: patients seen in the past for an identical period.	Forced expiratory volume (FEV1) and nocturnal pulse oximetry: Spirotel.	Spirometry data showed a significant improvement in mean annual Fev1 values for THC patients compared to the control group.
Hommerding et al. (2015)[15]	Effects of an educational intervention of physical activity for children and adolescents with cystic fibrosis: a	Randomized controlled trial. *N* = 34. Participant age: 7–20 years. Intervention	IG: aerobic physical exercise manual + stretching manual. They had to record when they performed aerobic exercise. They received phone calls	Forced expiratory volume (FEV1). Peak $\dot{V}$ O2. FEF forced expiratory Flow. $\dot{V}$ O2 oxygen uptake.	There were no physical changes between the two groups after the intervention.

(continued)

Table 2. Continued.

Author and year	Study title	Type of study	Intervention	Variables	Results
	randomized controlled trial.	time: 3 months.	every 2 weeks. CG: verbally he was given the notions of aerobic exercise.	BMI (body mass index)	
Rovedder et al. (2014)[17]	Exercise program in patients with cystic fibrosis: a randomized controlled trial.	Randomized clinical trial. *N* = 41. Participant age: 16 years and older. Intervention time: 3 months.	IG: home exercise program (aerobic and strength exercises) + telephone follow-up at 3 months. CG: physical therapist follow-up every two months.	Primary: quality of life. Secondary: 6 MIN. WALK TEST (6 mwt).	According to the results, no improvements were obtained in the quality of life of the patients who received the therapy.
Horck et al. (2017)[19]	Early detection of pulmonary exacerbations in children with cystic fibrosis by electronic home monitoring of symptoms and lung function.	*N* = 49. Participant age: 5–19 years. Intervention time: 1 year.	Using a home monitor, the children digitally recorded the presence and severity of respiratory symptoms (cough, sputum, and dyspnea).	Forced expiratory volume (FEV). Pulmonary exacerbations (PEX).	Electronic home monitoring of symptoms and FEV 1 can probably be used for early detection of PEX. Therefore, exacerbations could be predicted.
Gur et al. (2016)[20]	The use of telehealth (text messaging and video communications) in patients with cystic fibrosis: A pilot study.	Pilot study. *N* = 18 Participant age: >8 years Intervention time: 3 months.	IG: They received Skype video chats and WhatsApp messages from members of the CF multidisciplinary team. CG: without intervention.	Adherence and satisfaction.	The results show that the therapy in this study was feasible and acceptable. Knowledge, adherence, and patient satisfaction were similar in both groups.
Wood et al. (2019)[21]	A smartphone application for reporting symptoms in adults with cystic fibrosis improves the detection of exacerbations: results of a randomized controlled trial.	Randomized clinical trial. *N* = 60. Participant age: not specified. Intervention time: 12 months.	IG: use of an application. It included questions related to symptoms suggestive of an exacerbation. CG: usual care.	The primary outcome measure was the number of cycles and days of intravenous antibiotics.	Using an app reduced the time to detect respiratory exacerbations requiring antibiotics, however, it did not show a clear effect on the number of intravenous antibiotic courses.
Grzincich et al. (2010)[16]	Evaluation of a home telemonitoring service for adult patients with cystic	Pilot study. *N* = 60. Participant age: 19–44 years.	IG: telemonitoring with a recording device that allowed them to perform spirometry	Telemonitoring system: questionnaires	The results conclude that the pilot test was positive. Physicians reported

(continued)

Table 2. Continued.

Author and year	Study title	Type of study	Intervention	Variables	Results
	fibrosis: a pilot study.	Intervention time: It does not indicate.	tests, save data, and quality of life. The device stored the data and sent it to a data center. CG: conventional healthcare.	for patients and doctors.	that it was useful to monitor the health status of patients and that the workload in the CF center decreased. The difficulties that the patients show were the nocturnal saturimetry measurements.
Bathagate et al. (2021)[18]	Pilot RCT of a telehealth intervention to reduce symptoms of depression and anxiety in adults with cystic fibrosis.	Pilot study. $N = 31$. Participant age: over 18 years. Intervention time: 2 months.	IG: six sessions with option to a seventh telehealth session. CG: standard care.	Depression and anxiety. Coping self-efficacy and quality of life. Feasibility, acceptability, and satisfaction.	The intervention group presented better results with respect to the control group in depression, anxiety, coping and vitality.

health and more effective than no intervention in improving respiratory symptoms and quality of life in patients with cystic fibrosis. However, these results should be interpreted with caution due to differences in the intensity of the therapy and differences in effect sizes among the included studies. Telemedicine has also been suggested as a more motivating and enhancing form of exercise.

Adherence and satisfaction

Although we have not selected it as objectives, another aspect to highlight is the numerous articles that have evaluated adherence[14,19–21] and satisfaction with the non-conventional program.[13,16,18,20] Regarding adherence, a significant improvement was observed in three of the articles, one evaluated it by calculating the average percentage of adherence corresponding to the recommended frequency of transmissions.[14] Another article evaluated adherence by looking at the mean percentage of completed data compared to the total amount of data requested from all children and this was 77%, so more than half of the participants demonstrated good adherence (54%).[19] Adherence was also evaluated through the Treatment Adherence Questionnaire-CF (TAQ-CF).[21] The high level of adherence obtained is attributed to the low reporting load and the great use of today's technology.[21] Thanks to the recording of data in the mobile application of the intervention group, the detection time of exacerbations was also reduced.[17,21] In another of the articles, the level of adherence was recorded in the CF My Way10 program. There was no significant difference between both groups.[20] Finally, the protocol named in previous sections also studied adherence, calculating it by the relationship between the number of registered nebulizations compared to the number of prescribed inhalations; no results yet.[23]

With regard to patient satisfaction, all four studies have observed positive results. One of them examined satisfaction by means of a satisfaction questionnaire with telemedicine, and positive results were obtained.[13] Another of the studies used three types of questionnaires as follows: Questionnaire Q1 was carried out at the beginning of the study to see the satisfaction of the participants with the medical center in the previous 12 months. The Questionnaire Q2 was carried out at the end of the study to see the thinking of the patients regarding telemonitoring. The Questionnaire Q3 was made to the doctors to see their opinion on telemonitoring. It was found that for physicians, monitoring turned out to be useful and also decreased the workload in cystic fibrosis centers. Regarding the participants, 80% wanted to continue using this method in the future.[16] The third of the articles resulted in satisfaction through an ad hoc measure of eight articles and four open questions. The mean score for each item to measure acceptability and satisfaction was greater than 4 out of 5.[14,18] Finally, the last study evaluated satisfaction using a scale from 1 to 10 that patients had to fill in. A score of 8–10 was obtained.[20]

Two articles have been found, which also study through questionnaires, satisfaction with telehealth in patients with cystic fibrosis in the COVID-19 era. In one, the

participating population rated telehealth with very high values of convenience and satisfaction. In addition, they highlight that they enjoyed the necessary time in the appointments, that all their doubts were resolved and that all the desired disciplines were attended to; if they highlight the lack of sputum cultures.[24] In the other study, it is highlighted according to the responses to the questionnaires that the greatest benefit of telehealth is the saving of time by not having to go to the hospital and the ease with people who work or, in the case of children, not having to miss school. On the other hand, the lack of physical examinations also stands out among the results.[25]

One of our selected articles studies self-efficacy through the Stanford Self-Efficacy for Managing Chronic Disease 6-Item Scale for self-control of patients with cystic fibrosis. A significant increase in the scale score was obtained after the intervention of the experimental group.[12]

Type of telerehabilitation

The selected articles present different means for which the telerehabilitation has been carried out. Three of the 11 articles received telephone calls.[11,15,17] Two of the studies used an app a mobile phone application to improve self-efficacy in the self-management skills of people with CF.[12,21] Another article received Skype-based online video chats and WhatsApp messages from members of the CF multidisciplinary team.[20] He had between four and six Skype video chats and 22.45 WhatsApp messages.[20] The rest carried out telemedicine. Although it is true that telerehabilitation can be carried out through different methods, none of the articles talks about whether one method or another is better. It would be convenient to use one adapted to the average population in which it is used and to the capacity they have with the new technologies. In advanced ages, the most effective would surely be telephone calls, since it is the simplest and they would not have to handle applications.

A systematic review evaluated web portals as a means of telerehabilitation in patients with chronic diseases. The portals had several functions, including monitoring, follow-up, knowledge, health awareness, and more active participation by patients. Within the portals themselves, there were different means of communication, including text messages, forums, and videoconferences. Finally, for monitoring, most of the articles refer to the need for other digital tools. Therefore, finally, the common objective of telerehabilitation is to facilitate rehabilitation so that it becomes a daily lifestyle for patients.[26]

Future studies could assess the importance or relevance of using one type of telerehabilitation or another.

Telemedicine

One of the remaining five was monitored using a spirometer from home.[14] Another three carried out telemonitoring. One had a recording device that allowed them to perform spirometry tests, save oxygen saturation levels, and complete a symptom diary. The device stored the data and sent it to a data center.[16] Another monitored to measure the level of adherence and to measure the spirometric values that would be reported with access to the doctors. They also had the possibility of videoconferences.[13] In another case, a home monitor was used three times a week, recording the presence and severity of cough, sputum, and dyspnea, and uploading the data once a week.[19] Finally, the last of the articles used the Coping and Learning to Manage Stress with CF (CALM) intervention. It is a cognitive behavioral stress management (CBSM) telehealth intervention tailored for adults with cystic fibrosis.[18]

Measuring tools

As shown in this systematic review, the large number of measurement tools used in children and adults with cystic fibrosis can be appreciated to quantify the different variables. In other reviews, there is already talk of the little consensus that there is on which are the best tools to assess the results of the interventions.[27] Evidence was found in a study to trust spirometry, relating a poor FEV1 with progression to lung transplantation, a low quality of life, a higher risk of hospitalization, and pulmonary exacerbations. However, it has not yet been adopted as the preferred tool for quantifying lung function.[27] In this review, no consensus was found regarding the minimal clinically important difference (MCID) for FEV1.[27] They also summarize the different tools to measure quality of life (Table 4).[27]

Limitations and questions for future research

Within the limitations, we can find the small number of participants in the studies. In addition, the efficacy of telemedicine is being studied in all ages, and it would be convenient in future studies to review more focusing on children, adults, or older adults and to introduce studies with larger samples.

Conclusions

Most of the articles that have been examined did not show significant improvements in quality of life, forced expiratory volume, exercise capacity, and forced vital capacity in the experimental group versus the control group; if you detected an increase in physical activity in the intervention group. Good results have been obtained when it comes to detecting exacerbations, thanks to the continuous recording of data, the detection time of exacerbations has been shorter.

With regard to adherence and participant satisfaction with telemedicine, the vast majority of studies show significant results in favor of continuing with this method in the

future. It is noteworthy that the high level of adherence obtained is due to the low burden of reports, the great use of today's technology and the saving of time by not having to go to the hospital frequently. Physicians, for their part, point out that monitoring turned out to be useful and also reduced the workload in cystic fibrosis centers.

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ORCID iDs: María Jesús Vinolo-Gil https://orcid.org/0000-0003-4589-4442
Rocío Martín-Valero https://orcid.org/0000-0002-1664-3647

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Appendix

Appendix 1. Search strategy.

Databases	Total articles found	Search
PubMed	331	(Telemedicine) OR (Computing Methodologies/Therapeutic) OR (Computing Methodologies/Therapy) OR (Telerehabilitation) AND (Cystic Fibrosis). (Telerehabilitation AND telemedicine) AND cystic fibrosis. exercise programme AND cystic fibrosis.
Scopus	116	
Web of Science	87	
CINAHL	178	
PEDro	8	
Cochrane	1	