Shared Decision-Making Tool for Self-Management of Home Therapies for Patients With Cystic Fibrosis

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Objective: *Patients with cystic fibrosis (CF) undertake time-consuming programs of home therapies. Our objective was to develop a tool to help CF patients prioritize personal goals for some of these treatments. We describe the development and results of initial evaluation of this shared decision-making tool.* ***Methods:*** *Multicriteria decision-making method to develop a shared decision-making tool that integrates patient's values and perceptions of treatment impact on functionality/sense of well-being. Treatment efficacy data obtained through comprehensive review of English language literature and Cochrane reviews. Field study of 21 patients was performed to assess acceptability of the approach, understandability of the tool, and to determine whether there was sufficient patient-to-patient variability in treatment goals and patient preferences to make use of a personalized tool worthwhile.* ***Results:*** *Patients found the tool easy to understand and felt engaged as active participants in their care. The tool was responsive to variations in patient preferences. Priority scores were calculated (0–1.0 ± SD). Patients' most important treatment goals for improving lung health included improving breathing function (0.27 ± 0.11), improving functionality/sense of well-being (0.24 ± 0.13), preventing lung infection (0.21 ± 0.08), minimizing time to complete treatments (0.16 ± 0.12), and minimizing cost (0.11 ± 0.09).* ***Conclusions:*** *A shared decision-making tool that integrates patients' values and best evidence is feasible and could result in improved patient engagement in their own care.* ***Key words:*** *decision support tools; shared decision making; cystic fibrosis; analytic hierarchy process.* ***(MDM Policy & Practice XXXX;XX:1–14)***

Received 3 January 2017 from the Division of General Internal Medicine and the Center for Clinical Effectiveness (MHE) and Division of Pulmonary Medicine and Critical Care (EJK, KML, LPK, VMI, PMJ), University of Cincinnati, Cincinnati, Ohio; Pediatric Pulmonology, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio (LB, PMJ). Support for this study came from the Cystic Fibrosis Foundation Grant Number 15Q10 and NIH/NCATS Grant Number 1UL1TR001425-01. The funding sources had no role in the planning, design, or conduct of this study or the writing of this report. The findings and conclusions in this article do not necessarily reflect the view of the Foundation. Revision accepted for publication 15 May 2017.

The online appendixes for this article are available on the *Medical Decision Making Policy & Practice* Web site at http://journals.sagepub.com/home/mpp.

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DOI: 10.1177/2381468317715621

Cystic fibrosis (CF) is the most common life-threatening autosomal recessive disease among Caucasian populations, affecting roughly 1 in 2,000 live births. It has a major impact on both quality of life and survival, with a median life expectancy of 40.7 years.[1] Patients with CF are advised to undertake a comprehensive program of home therapies and activities including a variety of inhalation therapies, such as dornase alfa, antibiotics, and hypertonic saline.[2–4] Most patients are also advised to perform airway clearance, and to engage in regular physical activity and exercise.[5] To undertake all of these interventions and activities on a daily basis takes time and effort, and it may entail significant cost. One study found that CF home treatments took an average of 75 minutes each day (±57 minutes), and more when ill.[6]

Patients' preferences and values matter. In particular, involving patients in treatment decisions and providing opportunities for independence and autonomy is critical to success.[7] While developing

personalized approaches to improving adherence may be challenging and time consuming, such efforts have been shown to pay off.[8] Patient-centered strategies, such as shared decision making, improve adherence, at least in other chronic diseases.[9,10] In studies of CF patients, it has been shown that involving patients and their families in decision making is associated with positive perceptions about quality of care.[11]

Since patients may not be able to do everything every day, we have developed a program that can help them make informed decisions, prioritizing which of their treatments are most important to use on a regular and daily basis, and which best meet their goals when they are ill. Through this shared decision-making program, we can provide patients with information about the effectiveness of treatments and interventions while at the same time incorporating their input about what matters most to them.

METHODS

Development of Shared Decision-Making Tool

Our goal was to develop a tool that can 1) quickly and easily obtain patient values for prioritizing treatment goals and 2) facilitate shared decision making by showing patients and clinicians how their priorities affect home treatment decisions. We used steps described by the International Patient Decision Aids Collaboration to develop and perform initial evaluation of the CF Shared Decision-Making Tool (CF-SDM).[12,13]

Process Framework

We formed the following key groups to facilitate the development process:

1. *Project Management Group*—Provided executive control over the project and included (PMJ—project PI, Professor of Medicine, Division of Pulmonary, Critical Care, and Sleep Medicine, and Director of the University of Cincinnati's Adult Cystic Fibrosis Center; EJK—PhD-trained epidemiologist and program manager; and MHE—Professor of Medicine, Division of General Internal Medicine, and decision scientist experienced in the development of shared decision-making tools).
2. *Advisory Group*—Provided expert advice and ongoing feedback about content and proposed implementation and dissemination plans, and included (BT—Professor of Medicine and Pediatrics, Division of Pulmonary Medicine, with a research focus on CF and pulmonary innate immunity; LB—Assistant Professor of Pediatric Medicine, Division of Pulmonary Medicine, and Director of the CF Transition Program with Cincinnati Children's Hospital Medical Center; VI—Assistant Professor of Medicine, Division of Pulmonary, Critical Care, and Sleep Medicine, and Co-Director of the University of Cincinnati's Adult Cystic Fibrosis Program; KML—CF Social Worker; MM—CF Nurse Coordinator; TM—nutritionist; LH—respiratory therapist. In addition, 8 CF patients participated as reviewers of materials we developed and gave iterative feedback (AB, AH, CS, DD, GT, JE, RW, and SA).
3. *Scientific Reference Group*—Provided high-level expert opinion and was available for consultation at multiple points during the development process; played a key role in reviewing the synthesis of evidence (PMJ, BT, LB, and VI—see above; TB—Professor of Pediatrics, Division of Pulmonary Medicine, and Director of CFWELL).
4. *Technical Production Group*—Responsible for development of decision support tool prototype (MHE—see above; LK—computer programmer and application developer).

Major tasks were organized into a content specification phase (needs assessment, evidence synthesis, and consensus on evidence) and a design phase (initial prototype design, sandpit testing, usability testing, and field testing).

Needs Assessment

Assessment was done through a combination of informal discussions with patients during their clinical visits, and a formal questionnaire asking for open-ended input on the following:

1. Tell us about what you do on a daily basis to manage your CF. How much time, effort, and bother does it require?
2. Which of your daily CF treatments do you find sufficiently bothersome that you might not do them every day or at all? Tell us why (e.g., takes too much time, have to do them too often each day, costs too much)?
3. What medical outcomes or consequences of CF are you most concerned with (e.g., breathing function, lung infection)?

4. In what ways do you feel that home treatments (e.g., inhaled hypertonic saline, antibiotics or pulmozyme, airway clearance, exercise) benefit you in achieving your personal goals?

After soliciting input from our CF patients to determine what they considered the most important goals of home therapy, we developed the following list:

- Preventing lung infection
- Improving breathing function
- Improving functionality and feeling of well-being
- Minimizing time required each day to complete all treatments and interventions
- Minimizing cost

We next specified and developed consensus (Advisory and Scientific Reference Groups, and patients) regarding the necessary clinical content, the most important treatment goals for improving lung health, and the home treatment alternatives of interest, which were determined to be the following:

- Inhaled dornase alfa
- Inhaled antibiotics
- Inhaled hypertonic saline
- Airway clearance
- Exercise

Evidence Synthesis and Review of the Data Used in the Analytic Hierarchy Process (AHP) Model

The synthesis of evidence was facilitated by one of the principal investigators (MHE). Guidelines for the maintenance of lung health in CF patients recommend a wide variety of medications and home treatments.[2,4,14] We focused on interventions that are associated with poor adherence due to time and effort required on a daily basis and cost. We reviewed the English language medical literature and used Cochrane reviews to find the most up-to-date information regarding the efficacy of these treatments and interventions (see Table 1). Although some of the seminal studies are more than 10 years old, they are the best and still cited studies in the most recent Cochrane reviews.

Recombinant human DNase. For individuals with CF $\geq$ 6 years of age with moderate to severe lung disease, the CF Foundation strongly recommends the chronic use of dornase alfa to improve lung function and quality of life, and to reduce exacerbations.[2] This is a grade A recommendation for patients with moderate to severe disease and a grade B recommendation for those with mild disease. The 2010 Cochrane review reports only two studies examining the efficacy of dornase alfa compared with placebo in reducing pulmonary exacerbations.[15] The study by Fuchs and others reported a risk ratio of 0.81 (95% confidence interval [CI] = 0.61–1.06) in favor of dornase alfa at 6 months.[16] Quan and others reported on outcomes at 2 years with a risk ratio of 0.71 (95% CI = 0.49–1.02).[17] Given the longer follow-up, we used data from the study by Quan and others. The Cochrane review described a single study[16] reporting results as an absolute mean change in FEV_1, with a mean difference of 3.24% (95% CI = 1.03–5.45) in favor of dornase alfa over placebo at 2 years.[17]

Aerosolized antibiotics. Two antibiotics are currently approved by the Federal Drug Administration for use by inhalation in patients with CF, tobramycin and aztreonam lysine. Effectiveness of both agents are similar. In addition, other inhaled antibiotics, such as colistin, are used by some centers. We have used data on the efficacy of tobramycin for our CF-SDM, although the efficacy data for inhaled aztreonam is similar.[18] For individuals with CF $\geq$ 6 years of age with moderate to severe lung disease and Pseudomonas aeruginosa persistently present in airway cultures, the CF Foundation strongly recommends the chronic use of inhaled tobramycin to improve lung function and quality of life, and reduce exacerbations.[2] This is a grade A recommendation for patients with moderate to severe disease and a grade B recommendation for those with mild disease. The 2011 Cochrane review reports only two studies examining the efficacy of inhaled antibiotics compared with placebo in reducing pulmonary exacerbations.[19] The larger study by Chuchalin and others (161 patients) with longer follow-up (3–12 months) reported a risk ratio of 0.78 (95% CI = 0.59–1.03) in favor of inhaled tobramycin over placebo.[20] The Cochrane review reported a meta-analysis of three smaller studies with a total of 77 patients followed between 1 and 3 months.[21–23] The mean difference in FEV_1 was 9.48% (95% CI = 5.92–13.04).[19] We used data from the largest trial (520 patients), reporting a mean increase in FEV_1 (% predicted) of 10% in the tobramycin treated group compared to a 2% decrease in mean FEV_1 in the control group after 20 weeks ($P < 0.001$), resulting in a mean difference of 12%.[24]

Table 1 Efficacy of Home Therapies in Preventing Pulmonary Infection and Maintaining Pulmonary Function

		Pulmonary Infection (Probability of Infection)		Pulmonary Function (Mean Change in FEV_1 % Predicted)
Inhaled dornase	Treated	0.17 (17)[a]	Treated	+0.0004 (17)[b]
	No treatment	0.24	No treatment	−0.032
	Relative risk	0.71	Net improvement	+0.0324
Inhaled tobramycin	Treated	0.40 (20)[c]	Treated	+0.10 (24)[d]
	No treatment	0.51	No treatment	−0.02
	Relative risk	0.78	Net improvement	+0.12
Inhaled hypertonic saline	Treated	0.24 (25)[e]	Treated	0.0415 (27)
	No treatment	0.38	No treatment	
	Relative risk	0.63	Net improvement	
Airway clearance	Relative risk	1.0 (29)[f]	Treated	+0.1 (30)[g]
			No treatment	0.0
			Net improvement	+0.1
Exercise	Relative risk	1.0 (31, 32)[h]	Exercise	+0.0617 (33)[i]
			No exercise	−0.11
			Net improvement	+0.1717

Note: FEV_1 = forced expiratory volume at 1 second.
a. Mild disease, patient ages 6 to 10; 2-year follow-up.
b. Absolute mean change in FEV_1 at 2 years.
c. Follow-up between 3 and 12 months.
d. Change in FEV_1 at 20 weeks.
e. 48-week follow-up.
f. Based on Cochrane meta-analysis comparing oscillatory devices to conventional physiotherapy for outcome—days of hospitalization. No data available on pulmonary infections as outcome.
g. Best case estimate using non–statistically significant trend toward benefit over 1- to 6-month follow-up compared with conventional physiotherapy. Presumably, benefit compared with no treatment is no worse.
h. No studies have examined impact of exercise on lung infections.
i. Follow-up at 6 months.

Hypertonic saline inhalation. For individuals with CF $\geq$ 6 years of age the CF Foundation recommends the chronic use of inhaled hypertonic saline to improve lung function and quality of life and reduce exacerbations (grade B recommendation).[2] The largest multicentered study by Elkins and others enrolled 164 patients and compared twice-daily treatment with 7% saline compared with placebo (0.9% saline) over 48 weeks.[25] The hypertonic saline group had a significantly lower likelihood of pulmonary exacerbations, 24% versus 38% in the placebo group (P = 0.03), resulting in a risk ratio of 0.63 in favor of hypertonic saline. While this study did not find a significant improvement in lung function as measured by difference in FEV_1 between the two arms, a meta-analysis in the Cochrane review that included the Elkins study[25] and a smaller study by Eng and others[26] noted a net difference in FEV_1 of 4.15% (95% CI = 1.14–7.16).[27]

Airway clearance. There are numerous therapies for airway clearance.[28] We focused our review on oscillatory positive expiratory pressure devices. The guideline statement from the Pulmonary Therapies Committee is that airway clearance therapies are recommended for all patients with CF for clearance of sputum, maintenance of lung function, and improved quality of life (grade B recommendation).[5] No studies in the Cochrane review reported on the impact of oscillatory devices compared with either placebo or conventional chest physiotherapy on pulmonary exacerbations.[29] A similar lack of efficacy was noted in terms of days of hospitalization. We therefore assumed a relative risk of 1.0 for the impact of oscillatory airway clearance compared with no treatment. There was a nonsignificant trend toward benefit with regard to mean difference in lung function in a small study by Homnick and others.[30] Patients in the oscillatory device arm had a 10% higher FEV_1 than those in the conventional physiotherapy arm at a follow-up evaluation between 1 and 6 months (95% CI = −3.72 to 23.72).[30] Although this only

Table 2 Daily Time Required and Monthly Cost for Home Therapies and Activities

	Frequency	Time per Treatment (Minutes)	Total Daily Time (Minutes)	Monthly Cost ($)
Inhaled dornase	Once daily	25	25	2,800
Inhaled tobramycin	Twice daily	20	40	6,250
Inhaled hypertonic saline	Twice daily	20	40	85
Airway clearance	Three times daily	30	90	5
Exercise	Daily	30	30	30

supported a trend toward significance, we used this optimistic estimate in our model.

Exercise. We could find no reports describing the impact of exercise on pulmonary infections or exacerbations.[31] However, there is evidence that exercise significantly reduces the rate of decline in lung function in patients with CF.[32] The study by Kriemler and others had one of the longest periods of follow-up, with a mean difference of 17.17% (95% CI = 8.59–25.75) in favor of aerobic exercise versus no physical training noted at 6 months.[33]

Daily time required for home therapies. Total daily time required for each home therapy or activity was determined by multiplying the number of times per day the treatment or activity was performed by the time required for each treatment. The later information was obtained from a panel of patients and clinical experts who are members of our CF treatment team (see Table 2).

Cost. Monthly cost data for drugs (dornase alfa, inhaled tobramycin, and inhaled hypertonic saline) was obtained from www.GoodRx.com (see Table 2). The monthly cost for airway clearance devices was based on retail price for the Acapella Flutter Valve assuming a 10-month life span before replacement. Actual out-of-pocket costs to patients may vary widely based on insurance plans, copays, and deductibles.

Consensus on evidence. Iterative and ongoing discussions occurred at regular meetings of the Project Management group with the larger Advisory Group to discuss data obtained during the comprehensive literature review and develop consensus on parameter estimates to be used in the CF-SDM (see Tables 1 and 2).

Design Phase: Development of Decision Model

To build the computational engine for our CF-SDM, we used an approach frequently seen in the business world, known as the Analytic Hierarchy Process (AHP).[34] The AHP is one of a number of multicriteria decision-making methods. More recently, it has been applied to address medical decision making.[35] In short, there are three stages in the development of an AHP model. In the first stage, the problem is described through a hierarchy of multilevel decision elements (see Figure 1). We used what we learned from our patients during the needs assessment to structure the hierarchy. The second stage involves making pairwise comparisons of elements within each level of the hierarchy with respect to their importance or impact on elements above them in the hierarchy (see Figure 2). These pairwise comparisons allow relative weights to be calculated prioritizing each element in each level of the hierarchy. The last step involves applying these weights to calculate the relative score for each of the decision alternatives. While subjective responses, using pairwise comparisons, are frequently used to calculate weights for all elements in the hierarchy, we used quantitative data on treatment efficacy to calculate weights wherever possible, along with actual costs and time estimates. These later weights are the same for all personalized analyses done by the AHP, while the weights representing each patient's personal values for treatment goals vary from patient to patient.

Thus, we use the AHP model in real-time with individual patients to facilitate a shared decision-making discussion. Patients are given a CF-SDM booklet (see the online appendix) that provides background information and context for using the tool, along with a discussion of the goals of treatment, the home treatment and intervention alternatives, the purpose of the shared decision-making tool, and sample results for a hypothetical patient. The next section of the booklet is titled ''Gathering Information About You,'' and it presents patients with a series of 10 pairwise comparisons (see Figure 2) of treatment goals. Patients provide their relative priorities and preferences for treatment goals by circling the appropriate numbers on these scales.

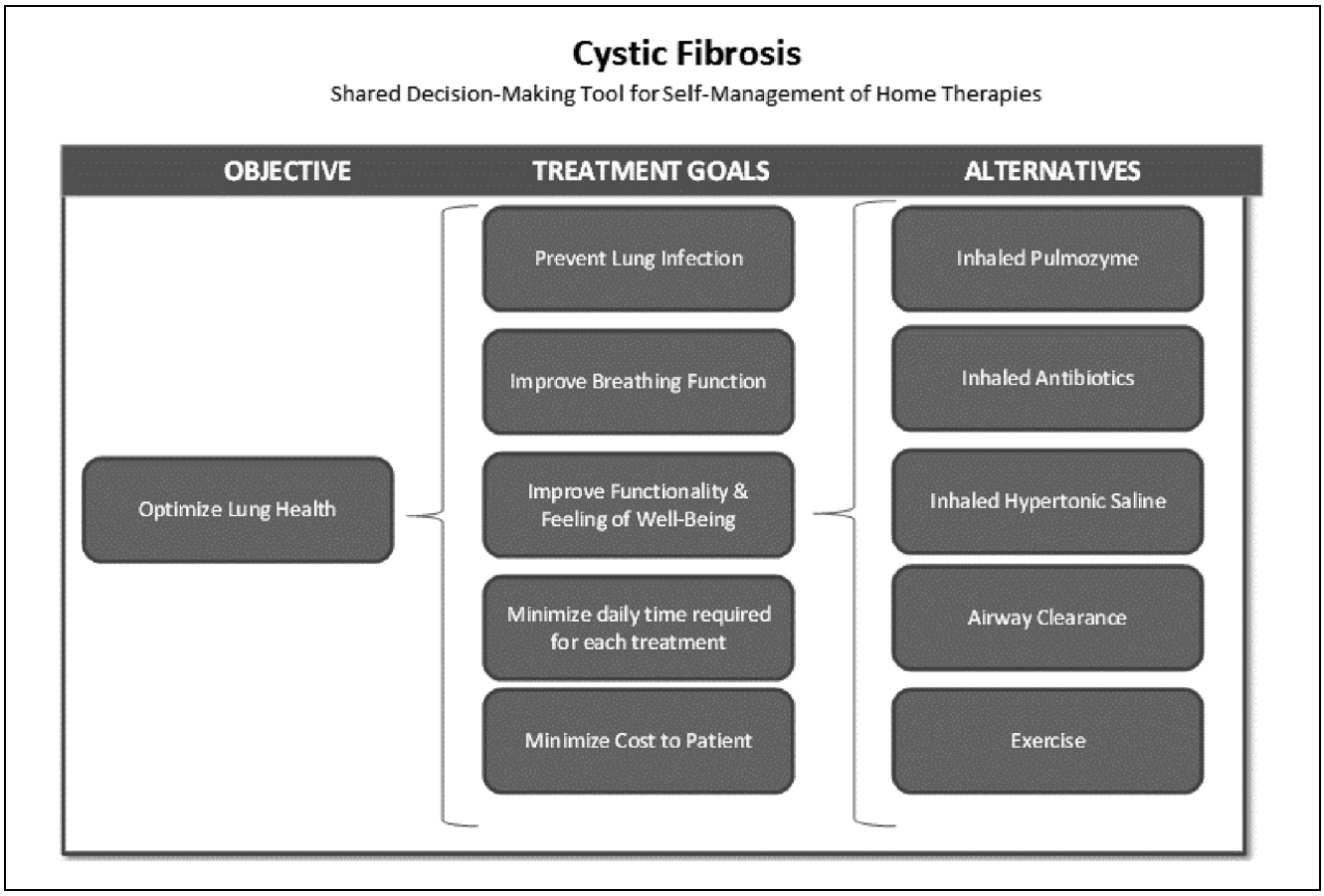

Figure 1 Analytic hierarchy model for patients with cystic fibrosis (CF). Analytic hierarchy structure for shared decision making about self-management of home therapies for patients with CF. The top level objective is to optimize lung health. The second level of the hierarchy illustrates the treatment goals that lead to optimizing lung health, while the third level of the hierarchy depicts treatment alternatives that differentially impact the treatment goals in the second level of the hierarchy.

Patients are next asked for their personal assessments of the impact on functionality and sense of well-being they have experienced with the treatments and interventions under consideration. This information is provided once more, by circling the appropriate number on each of another 10 pairs of comparisons. In the current prototype of the CF-SDM undergoing field testing, this information is then manually entered by a study coordinator into the AHP calculational model. The AHP model was built using a generic Microsoft Excel spreadsheet template for constructing AHP models provided by Padilla-Garrido and colleagues.[36] This can be downloaded free from journal *Medical Decision Making* Web page.[37]

The results of the AHP model are then copied into a personalized report template that shows the calculated priority score for each of the five different home therapies and interventions, based on both quantitative data from the medical literature regarding treatment efficacy and information patients have provided regarding their priorities for treatment goals (see Figure 3 for a sample report and the appendix for full personalized report booklet). Our future goal is to develop a self-contained computer application that will incorporate the functionality of the current CF-SDM paper and spreadsheet prototype and allow for patient education, collection of personalized value and preference information, calculation of the AHP model's personalized results for individual patients, and reporting of those results in a single seamless package.

While the typical approach to the AHP would involve asking patients to provide their subjective

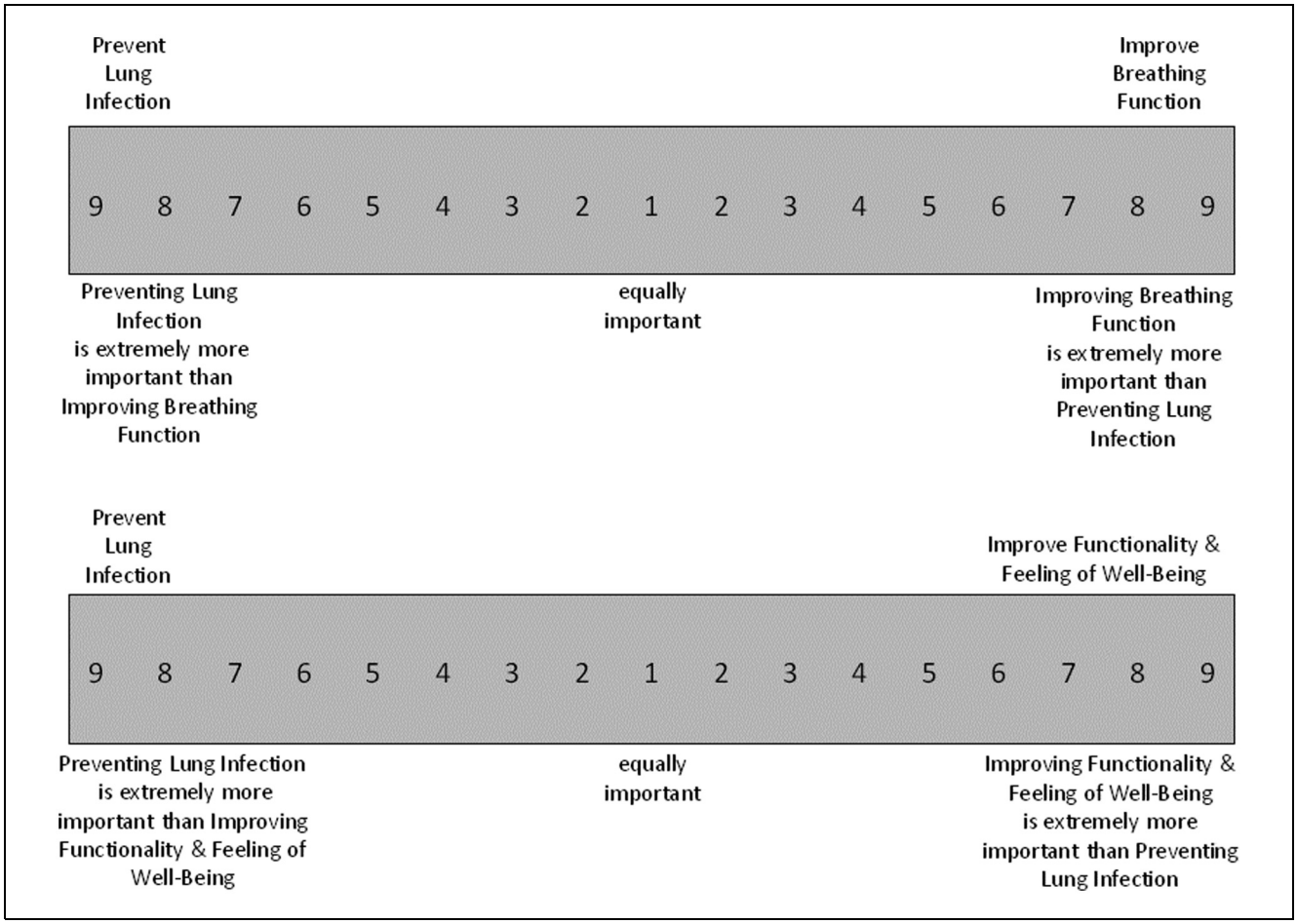

Figure 2 Example of pairwise comparisons used to help patients prioritize treatment goals. In the top panel, the importance of preventing lung infection compared with improving breathing function is assessed on a 9-point scale. If a patient felt that preventing lung infection was extremely more important than improving breathing function, they would circle the number 9 at the far left of the scale. If they felt that improving breathing function was extremely more important than preventing lung infection they would circle the number 9 at the far right of the scale. If they felt these two treatment goals were equally important they would circle the number 1 in the middle of the scale. The bottom panel assesses the relative importance preventing lung infection versus improving functionality and feeling of well-being. A total of 10 pairwise comparisons are assessed to determine the relative importance of each of the five treatment goals.

judgements regarding the pairwise comparison of all elements at all levels of the hierarchy, resulting in 50 such comparisons in our model, we tried to minimize the cognitive burden by ''hard-wiring'' quantitative data when available from either clinical trials in the medical literature or expert opinion from our CF clinical team, as described above. We obtained data from the medical literature regarding the effectiveness of the treatments considered. We also compiled information for the average retail cost of treatments (www.goodrx.com), and the time required each day to complete them from our clinical experts. Thus, the CF-SDM only requests individual patient's input regarding what patients are most expert in providing 1) the relative importance of each home treatment and health goals and 2) the impact of each treatment or health intervention on improving their *functionality and feeling of well-being*.

Sandpit Testing

In the early design phase, we experimented with alternative graphical approaches for requesting patients' priorities for treatment goals and for presenting data to patients. We ultimately developed a

pamphlet that explained the purpose of the CF-SDM, the goals of treatment, and treatment alternatives (see the appendix). The pamphlet also was designed to gather information from patients about their personal preferences and values for the five health treatment goals and their opinions regarding the impact of each treatment and intervention on improving their functionality and feeling of well-being. The latter set of questions regarding patients' view of the impact of treatments also captures in a holistic manner, a number of more subjective components, including patients' preferences for the five treatments.

We developed a separate pamphlet that contained a personalized report for each patient, showing graphics for the relative importance of each treatment goal based upon their responses to the series of pairwise comparisons they were asked to complete, and their personal prioritization of home treatments based on application of the AHP (see Figure 3 for graphics and the appendix for full pamphlet).

Usability Testing

We tested prototypes of the CF-SDM tool in meetings with clinicians involved in the care of CF patients (pulmonologists, dieticians, nurse practitioners, respiratory therapists) and a series of CF patients. Using an iterative process,[12] we presented the pamphlets to patients and physicians, determined what they had difficulty understanding, and obtained their feedback about what we could improve or add. We then updated and improved the pamphlets. We went through several iterations of this process until few additional comments or requests were made.

Field Testing

We next field tested the CF-SDM tool on a sample of 21 patients age 20 to 66 years, mean age 31.4 years, 11 female (52%) and 10 male (48%). Table 3 describes the patients' characteristics. This was a convenience sample; patients were approached as part of our quality improvement project for CF care while they already were at the clinic for a scheduled visit. Patients were asked to 1) assess ease of use and understandability, 2) get general reactions to use of the tool, and 3) assess the degree of variability in patient-specific responses for relative importance of treatment goals, and patients' assessment of the efficacy of each treatment on their functionality and feeling of well-being (see the appendix for full text of Understandability and Usability Questionnaire).

RESULTS

Understandability and Usability

Twenty-one patients filled out the questionnaire developed to assess understandability and ease of use of the CF-SDM (see the appendix for complete questionnaire). The items assessed whether patients found the tool helpful in clarifying their personal values, improving their preparedness to discuss home treatment options with their physicians, improving their sense of engagement in decision making about their home therapies, and whether they felt the personalized report and recommendations accurately reflected their preferences and values. Virtually all patients agreed or strongly agreed with all 12 structured items. We also solicited unstructured comments about how they planned to use the information contained in their personal reports, what they liked or did not like about the booklets, and whether they would suggest adding other information (see Table 4). Reactions were uniformly positive. Most patients commented that the booklet was well laid out, that it was easy to understand and not too ''verbose'' or ''overwhelming.'' In general, patients reported not requiring more than 5 to 10 minutes to review the booklets.

Patients' Values and Preferences for Treatment Goals and Assessment of Treatment Impact on Functionality and Sense of Well-Being

As shown in Appendix Figure 1, there was substantial variability in patients' assessments of the pairwise comparisons of treatment goals. The same was true of patients' pairwise assessments regarding which treatments provided greater functionality and sense of well-being (see Appendix Figure 2).

Patient-Specific Results of the AHP

Figure 4 summarizes results for the patient-specific prioritization of home therapies and interventions. Exercise was rated highest on average (27 $\pm$ 4%) while the other four home treatments had similar average priority scores. However, it is

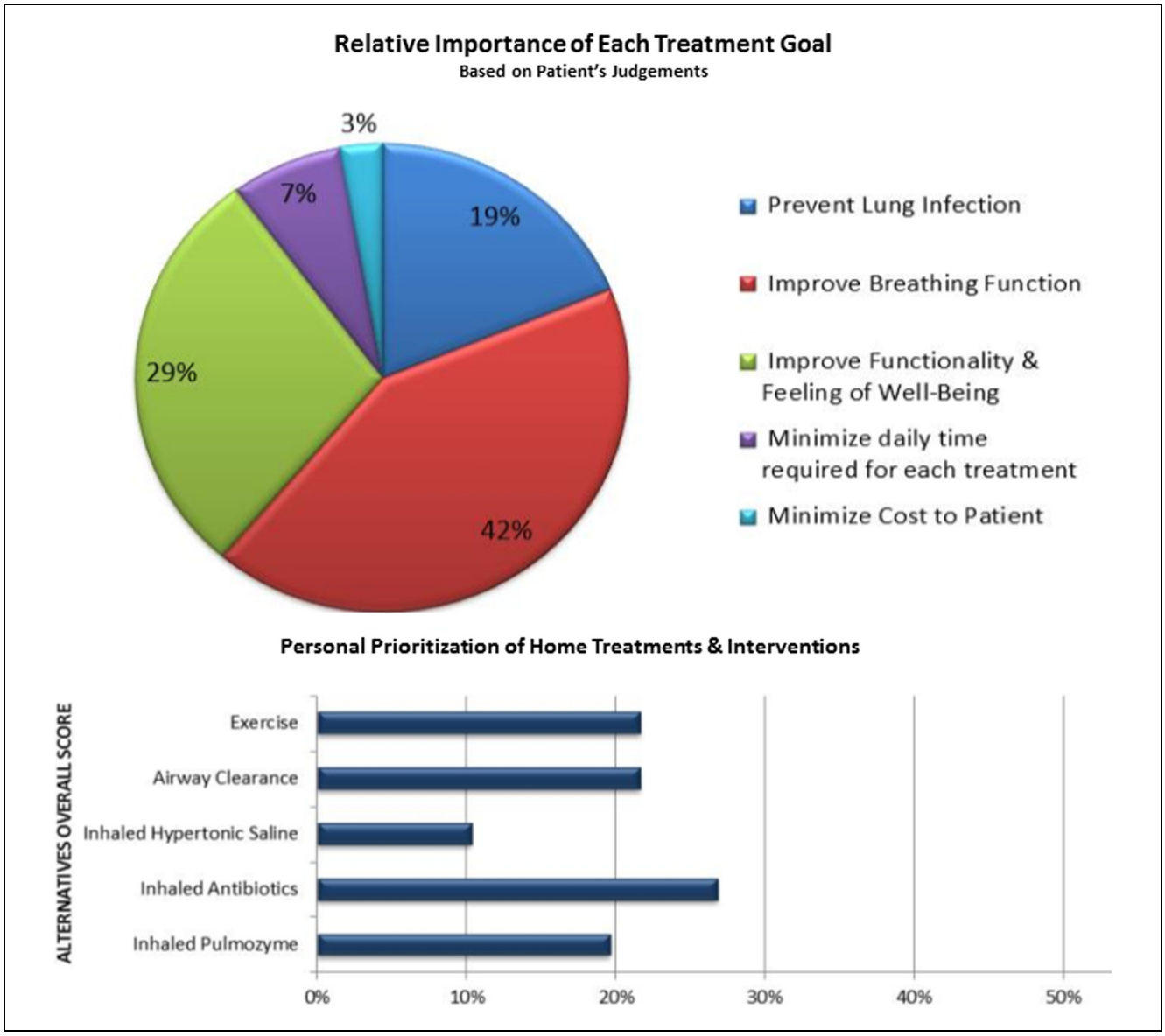

Figure 3 Personalized patient report. Example of graphics in personalized patient report showing the relative importance of each treatment goal (upper panel) and a prioritized listing of home treatments and interventions (lower panel). For this patient, improving breathing function was the most important treatment goal, followed by improving functionality and feeling of well-being, and preventing lung infection. In the bottom panel, the combination of this patient's values and preferences along with quantitative data regarding treatment efficacy, cost, and daily time required for each intervention result in a personalized ranking of treatment priorities such that adhering to inhaled antibiotics, performing airway clearance, and exercising on a regular basis are most important. If this patient found they could not adhere to every treatment and intervention on a daily basis or needed to add therapies due to early symptoms of exacerbation, inhaled hypertonic saline would have the lowest priority and personal yield, and perhaps could be a treatment that occasionally could be missed if absolutely necessary.

important to note that for any given patient, the pattern of priority scores was very different as was the pattern of individual patient's prioritization of treatment goals.

Appendix Figure 3 shows the relative importance of each of the five treatment goals for the 21 patients in our pilot study on a scale that runs from zero to 100%, along with the mean and standard deviation.

Table 3 Patient Characteristics

	n	SD	Range
Age (years)	31.4	25	20–66
Gender (female/male)	11/10	52%/48%	
BMI (kg/m^2)	25.3	8.4	16.8–52.3
FEV_1 at visit (%)	59	30	19–108
Best FEV_1 in last year (%)	66	28	19–11
Difference between current and best FEV_1	−7	10	−45 to 0
Hospitalizations in past year	1.4	1.8	0–5
Pulmonary infections treated in past year	2.5	2.1	0–7

Note: BMI = body mass index; FEV_1 = forced expiratory volume at 1 second.

Table 4 Sample of Qualitative Responses About the Shared Decision-Making Tool

"I feel that this booklet will be helpful in determining for new patients the most important aspects of their individual care."
"I think it's all a cycle or a puzzle. All pieces are important to achieve your best version of your health."
"I like how my doctor see's the report to better understand what is important to me."
"Will help me realize what I may not be focusing on and makes me aware of the changes I can make."
"I liked that it was a very quick process."
"Regarding the pairwise comparison of improved functionality and feeling of well-being versus minimizing total daily time required for each treatment—It's different since I don't work anymore, but time is important if you have a life and are busy!"
"Well, I definitely plan to exercise more, as this is just one more source telling me how important exercise is."
"I like that it's not too verbose. There's not too much information so that people get the way the booklet is laid out. It does a good job of explaining itself without being overwhelming and everything is just very succinct."
"I feel it is well designed."
"I found it detailed and overall good!"
"The graphs were great visuals. Really liked how 'feeling of well-being' was included as a concern."

For instance, the average importance of minimizing cost is 11% with a standard deviation of 9%. Improving breathing function had the highest relative importance of all five treatment goals (27 ± 11%), followed closely by improving functionality and feeling of well-being (24 ± 13%) and preventing lung infection (21 ± 8%). Minimizing cost was the least important treatment goal on average.

Sensitivity Analyses

We tested the sensitivity of patient-specific results and reports by examining the lower and upper 95% confidence limits describing the efficacy of each of the five home therapies and activities. While priority scores changed as expected across sensitivity analyses for the efficacy of the five home therapies, the rank ordering of priority scores was little affected. Thus, the overall recommendations of the shared decision-making tool are robust across known variation in values for these parameters.

DISCUSSION

As treatments improve and patients with inherited disorders such as CF thrive and survive into adulthood, engaging these patients in decisions about their own health, health care, and wellness is particularly important. As children and adolescents, their doctors and their parents dictate most therapeutic decisions and oversee their home activities. These treatments and activities are time consuming, at times complex, and place a significant burden on patients and their families. Furthermore, adherence is reported to decrease as children transition into adulthood.[38] Numerous barriers to treatment adherence have been described in adolescents and young adults with CF.[38–41] Treatment burden,

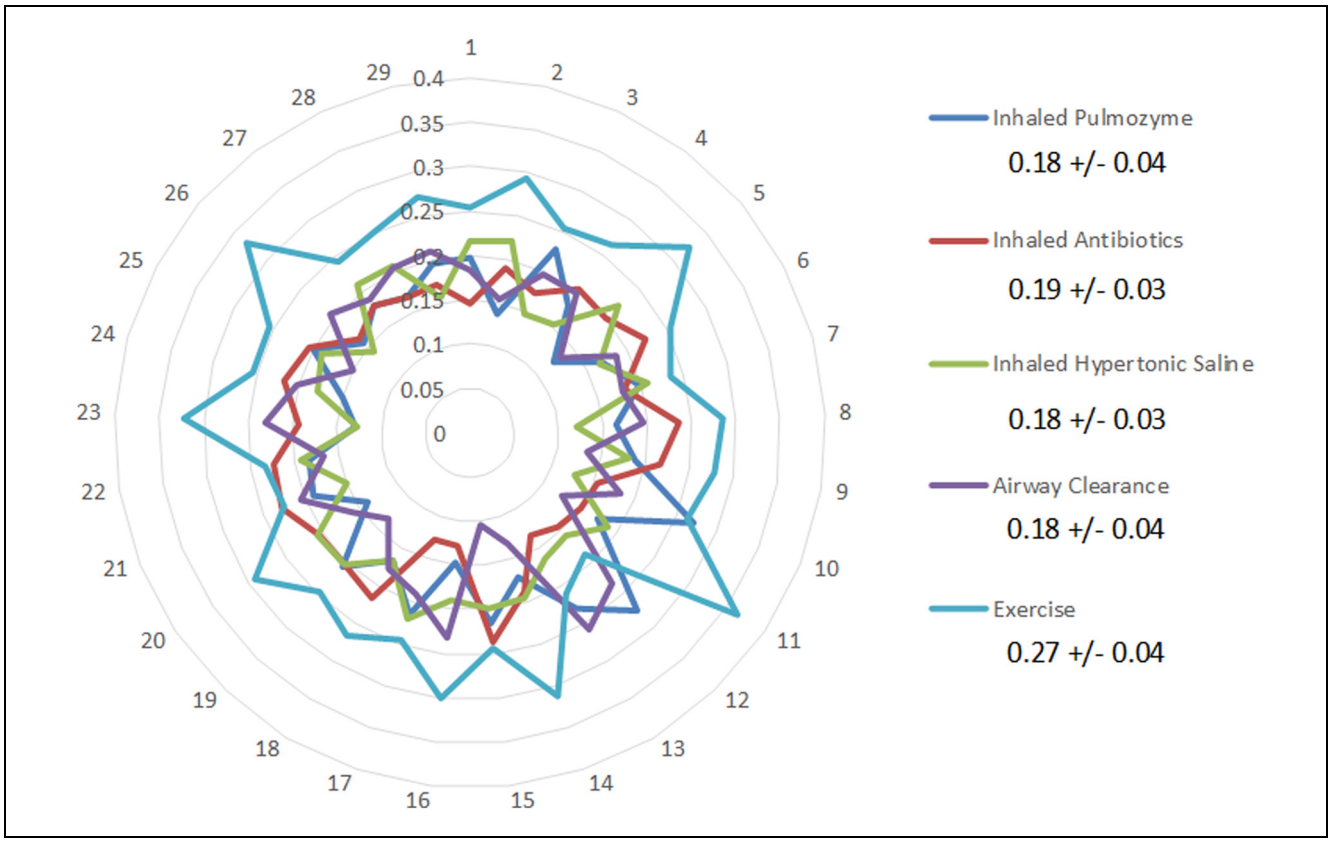

Figure 4 Relative importance of treatment goals. Radar chart showing the AHP-computed results for each patient's priority score for the five different home therapies and interventions. These are the final results shared with each patient. The priority scores must sum to 100% across the five treatments for each patient. Thus, the scores reflect the relative importance of each home therapy or activity for each patient. Each colored band corresponds to one of the five home therapies. The numbers around the circle indicate each patient in the study. The mean priority score across the sample of 21 patients is shown to the right, along with the standard deviation for each treatment.

forgetfulness, lack of perceived benefit, and rebellion from the yoke of parents' mandate have been identified as common barriers and explanations for poor adherence.[38] However, facilitators of adherence also have been identified for these patients transitioning from childhood to adulthood. Feedback of health information, such as pulmonary function test results, and patient-centered counseling that acknowledges the burden of home therapies and seeks to understand the patient's lifestyle and health goals, offering an opportunity for collaborative problem solving have been identified as powerful facilitators of adherence. Outcome expectancies and confidence in the efficacy of prescribed treatments also have been shown to affect self-management practices. Addressing these perceptions by sharing best evidence from clinical studies while also acknowledging patients' own experiences regarding the relative impact of home therapies on their functionality and feeling of well-being may be a way to dismantle some of these barriers to adherence.

To our knowledge, the only published example of a decision aid for patients with CF was a study by Vandemheen and colleagues focused on decisions regarding lung transplantation in CF patients.[42] The Cochrane Library is planning a systematic review of "interventions for promoting participation in shared decision-making for children and adolescents with cystic fibrosis," but to date only a protocol has been published.[43]

The many components of CF care can make shared decision-making complicated. The present approach to CF care has been to continually add

new therapies onto the existing regimen, often with limited input from patients. The CF-SDM provides a framework for patients and caregivers to collaborate in the development of a care plan that incorporates patient opinions and preferences.

Our field study of 21 patients with cystic fibrosis found that patients uniformly believed the shared decision-making exercise helped them develop personalized priorities for home therapies and activities. Use of the tool helped them clarify their personal values for the relative importance of home treatment goals and helped them feel better prepared to discuss home treatment options with their doctors. Perhaps most important, using the CF-SDM made them feel that they were contributing to making decisions in their care.

Of interest, patients differed significantly in what they identified as the most important goals of their home treatment regimens. They also reported varied perceptions regarding the relative impact of these home therapies on their functionality and sense of well-being. This argues all the more for the development of shared decision-making tools, like the CF-SDM, that can leverage this patient-to-patient variability in values and preferences.

Users must consider several factors when adopting the tool. There is a lack of newer efficacy data and limited data on drug interactions. Certainly the benefits of some therapies such as inhaled tobramycin may have changed over time. Although these data may be dated, they remain the current basis for CF care guidelines.[44] Patients were allowed to interpret the terms of the CF-SDM for themselves with limited input from the care team. After the surveys were developed, patients reported that they understood ''improve breathing function'' to mean improve pulmonary function testing, primarily FEV_1 but also included, to a lesser degree, difficulty breathing. ''Prevent lung infection'' was interpreted to mean avoiding treatment with antibiotics for a pulmonary exacerbation.

In reviewing the therapies and intervention alternatives, patients were asked about the perceived efficacy of these therapies. In the ranking of these options, patients may have been influenced by other factors such as the perceived inconvenience or taste of medication. The relevance of efficacy for an individual patient is often difficult to assess. It was our intention that they rank therapies based on personal experiences. For example, patients should rank a drug low if they have side effects such as wheeze or may rank inhaled antibiotics low if they do not grow Pseudomonas.

This CF-SDM was developed for use in adults with CF. This population should have input into their care and have opinions about the care they want. The tool may not be directly applicable to a pediatric population. Pediatricians may feel obligated to make recommendations based on what is felt to be best for the patient, and may be less inclined to incorporate the opinions of a child or parent/caregiver. Furthermore, the CF-SDM options may be less appropriate for smaller children who may not be able to perform pulmonary function testing or be candidates for inhaled antibiotics.

Finally, the current CF-SDM prototype is paper-based and uses a spreadsheet template to perform the personalized AHP calculations used to generate patient's personalized report booklets. Our future goal is to incorporate the full functionality of the prototype into a self-contained application that can be implemented on a computer tablet platform. A tablet running the application could be given to the patient to use while he/she is in the waiting room prior to a visit, and then used during the clinical visit to facilitate a shared decision-making conversation about home therapies.

Our next step is to perform a randomized clinical trial to evaluate the impact of a shared decision-making visit facilitated by the fully computerized CF-SDM versus ''usual care.'' Major outcomes of interest will include measures of decision quality, such as 1) decisional conflict,[45] confidence in decision,[46] and satisfaction with decision[47]; 2) patient knowledge regarding benefits of treatments; 3) quality of the therapeutic alliance between patients and the multidisciplinary care team[48]; 4) adherence to treatment; and 5) clinical outcomes.

Practice Implications

The goal of this initial study first was to demonstrate that a shared decision-making tool for patients with CF was usable, easy to understand, and improved patients' perceptions of engagement by promoting collaboration with their care team to develop a personalized care plan. The typical goals of shared decision making include improving confidence in the decision-making process, and increasing the sense of collaboration and trust in the care process. A shared decision-making tool that integrates patient's values and best evidence is feasible and could result in improved patient engagement in their own care. While we describe an application to patients with CF, the shared decision-making tool

could be extended to other aspects of CF care and management and to other chronic conditions, ultimately modifying the way we practice healthcare.

Conclusion

The future challenge of CF care will be to develop personalized care plans that are based on standards of care while still incorporating patient preferences and goals without compromising outcomes. Whether such engagement will result in improved adherence to their home treatment regimen and ultimately in improved clinical outcomes is an interesting and important question that we hope to answer shortly through a randomized clinical trial.

ACKNOWLEDGMENTS

We thank Jim Dolan, MD, for sharing his expertise in use of the analytic hierarchy process in the development of our AHP model. We also wish to thank Melenie Meyers and Traci Major for their insights and suggestions and assistance in getting patient feedback on the shared decision-making tool as we iterated through its early development.

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