Medical Mysteries: Investigating Improved CF-ILD Care Through Patient Empowerment

Live Symposium Outcomes Report *(held in conjunction with AARC 2020)*

BI Grant ID: IME_2020_00001820

American Association for Respiratory Care

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Overview

Activity Description: This webinar, held in conjunction with the AARC 2020 Virtual Congress, featured Gregory Cosgrove, MD and Sharon Tucker, RRT, who explored the role of RRTs in identifying misdiagnosed or at-risk patients with Chronic-Fibrosing Interstitial Lung Disease (CF-ILD). With the help of a patient and PFF Ambassador Emeritus, Dave Sherry, the panel discussed pulmonary function monitoring, pulmonary rehabilitation, and examined patient needs to maximize QoL through comprehensive care plans.

Symposium Date: Friday, November 20, 2020

Credit: 1.5 Continuing Respiratory Care Education (CRCE) contact hours

Sponsored by: The Academy for Continued Healthcare Learning (ACHL) in collaboration with the American Association for Respiratory Care (AARC)

Supported by: An educational grant from Boehringer Ingelheim Pharmaceuticals

Intended Audience: ID specialists, hospitalists, pharmacists and other healthcare professionals who care for patients with serious infections

Enduring Activity: An enduring CRCE highlights activity capturing key content from the live meeting will release in early January 2021 and will be hosted on AARC University (online learning portal with over 52,000 RRT members)

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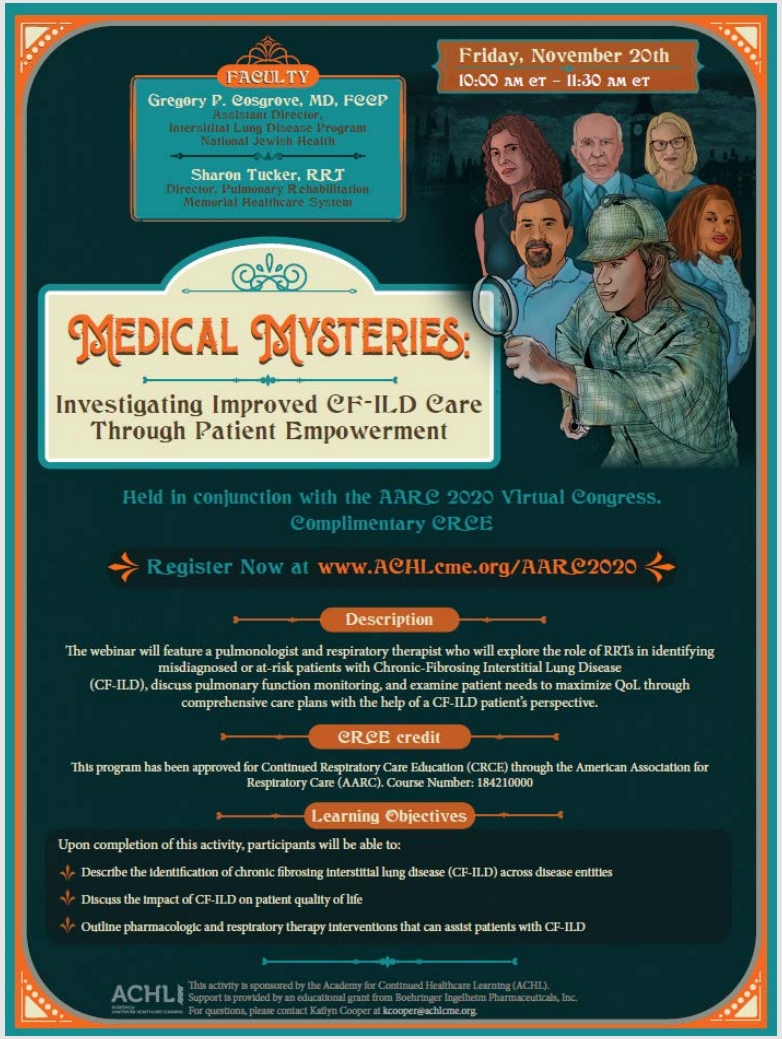
Activity Visuals

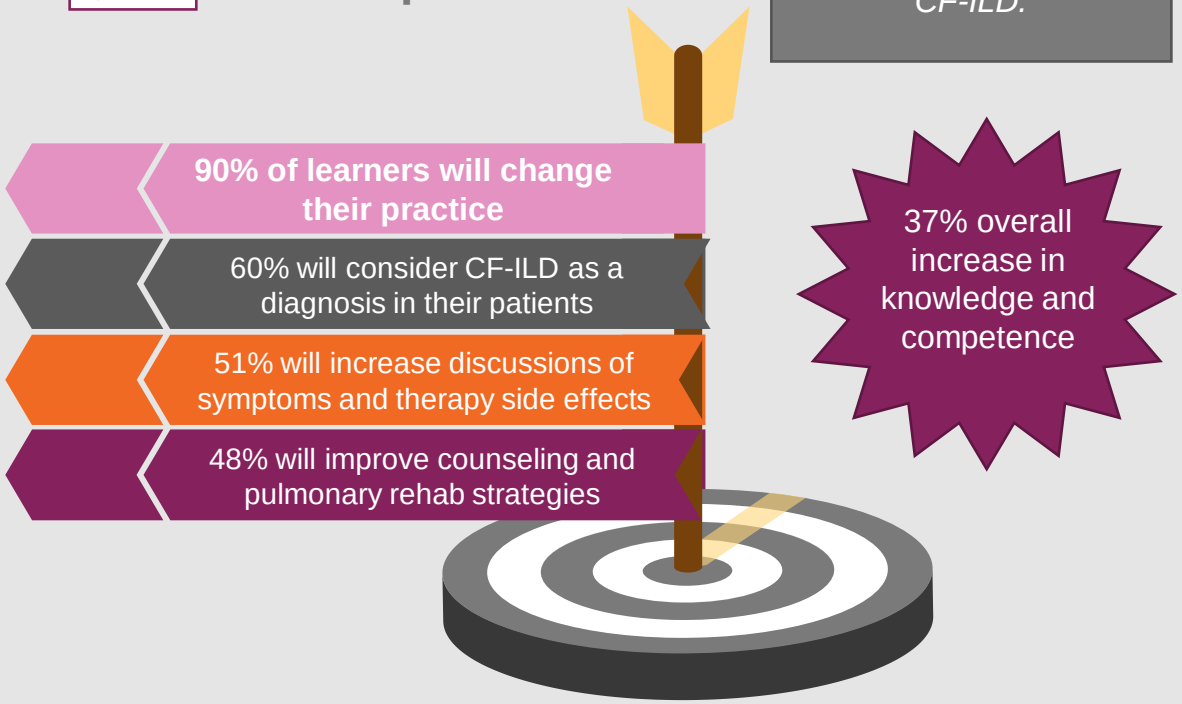


Executive Summary

Participants	Certificates Issued
140	67

95% Respiratory Therapists

Following the activity, nearly two-thirds of learners supposed they saw 1-5 patients in the past year who may been misdiagnosed or not yet properly diagnosed with CF-ILD.



37% overall increase in knowledge and competence

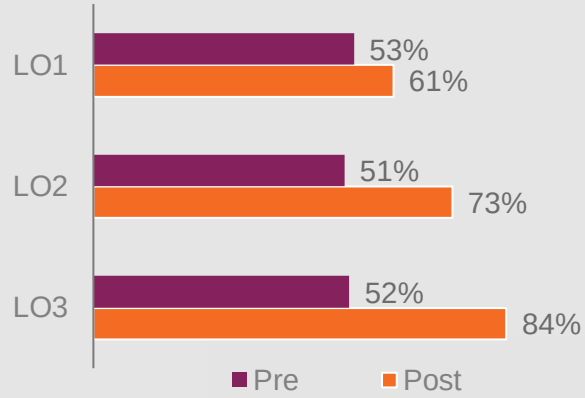
An **effect size** of 0.68 indicates that participating HCPs are now ~42.04% more knowledgeable of the content assessed than prior to participating in this symposium.

Changes will impact 125 to more than 397 patients with CF-ILD each month.

There was a 318% increase post-activity in learners who self-identified as “very confident” with discussing available therapy and supportive care of patients with CF-ILD.

Learning Gains

Describe the identification of progressive-fibrosing interstitial lung disease (CF-ILD) across disease entities
Outline pharmacologic and respiratory therapy interventions that can assist patients with CF-ILD
Discuss the impact of CF-ILD on patient quality of life



Future Education Opportunities

- Distinguishing CF-ILD from other respiratory conditions
- Diseases that may be associated with a progressive phenotype

- Patient impacts of CF-ILD therapy
- Personalizing CF-ILD comprehensive care plans

Learning Objectives & Overall Evaluation

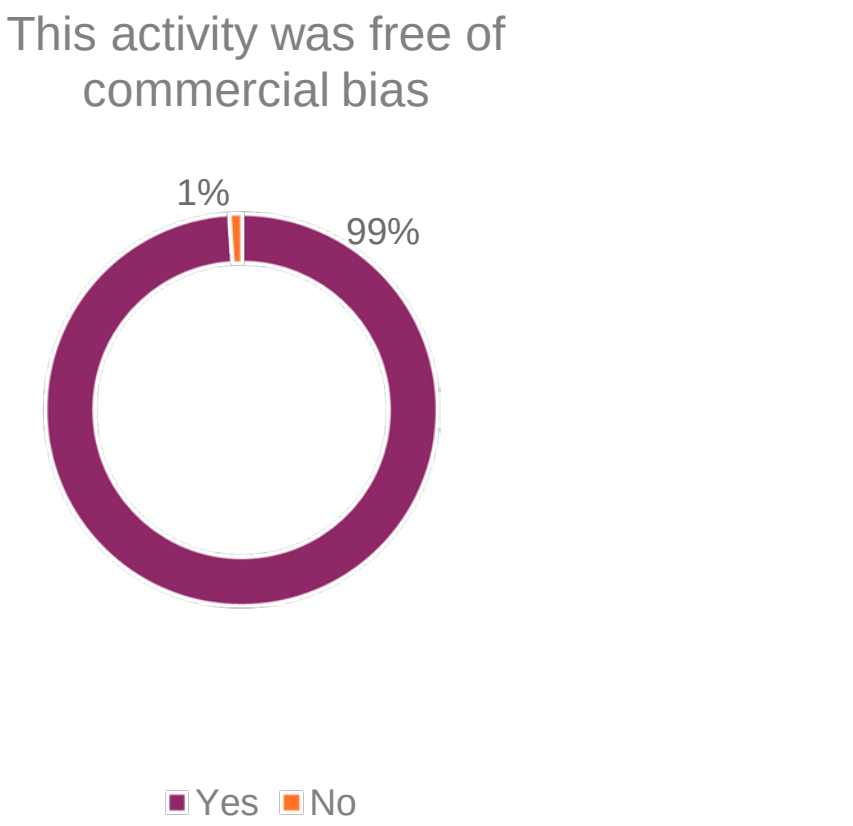
Please rate the following objectives to indicate if you are better able to: **Rating scale: 4=Strongly Agree; 1=Strongly Disagree**	**Analysis of Respondents**
Describe the identification of progressive-fibrosing interstitial lung disease (CF-ILD) across disease entities	3.69
Outline pharmacologic and respiratory therapy interventions that can assist patients with CF-ILD	3.70
Discuss the impact of CF-ILD on patient quality of life	3.73

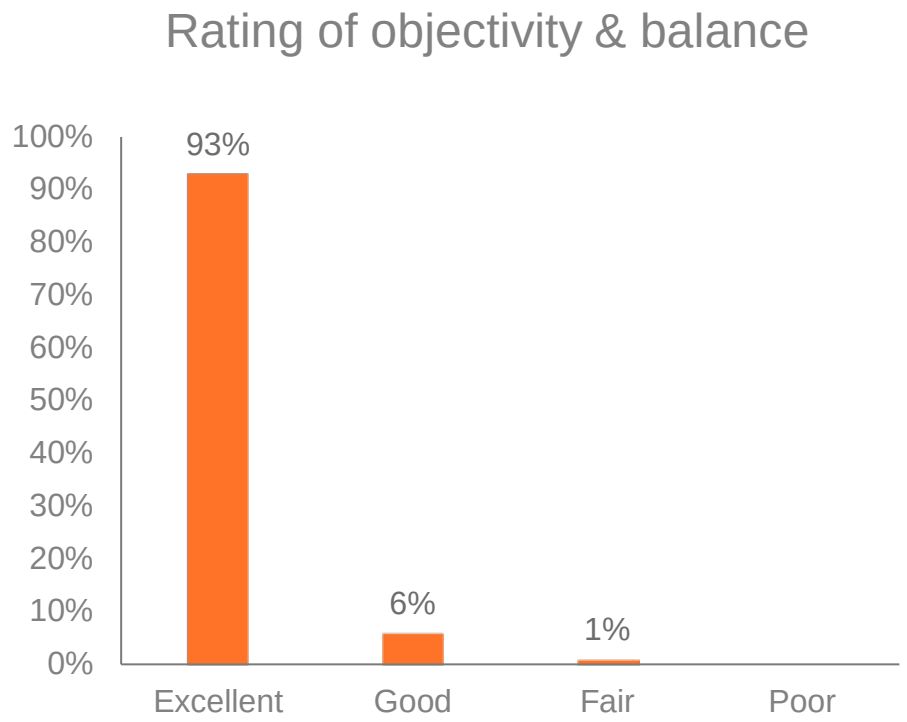
99% of learners strongly agree or agree that all learning objectives were met, with an average rating of 3.71/4.0

Please evaluate the following criteria: **Rating scale: 4=Excellent; 1=Poor**	**Analysis of Respondents**
Quality of educational content	3.91
The presenters' ability to effectively convey the subject matter	3.93
Inclusion of patient panelist and value to education	3.93

N=67

Objectivity & Balance





Activity was perceived as objective, balanced and non-biased.

N=67

Interactivity Assessments

Total responses (n) may fluctuate as all feedback and survey questions were optional.

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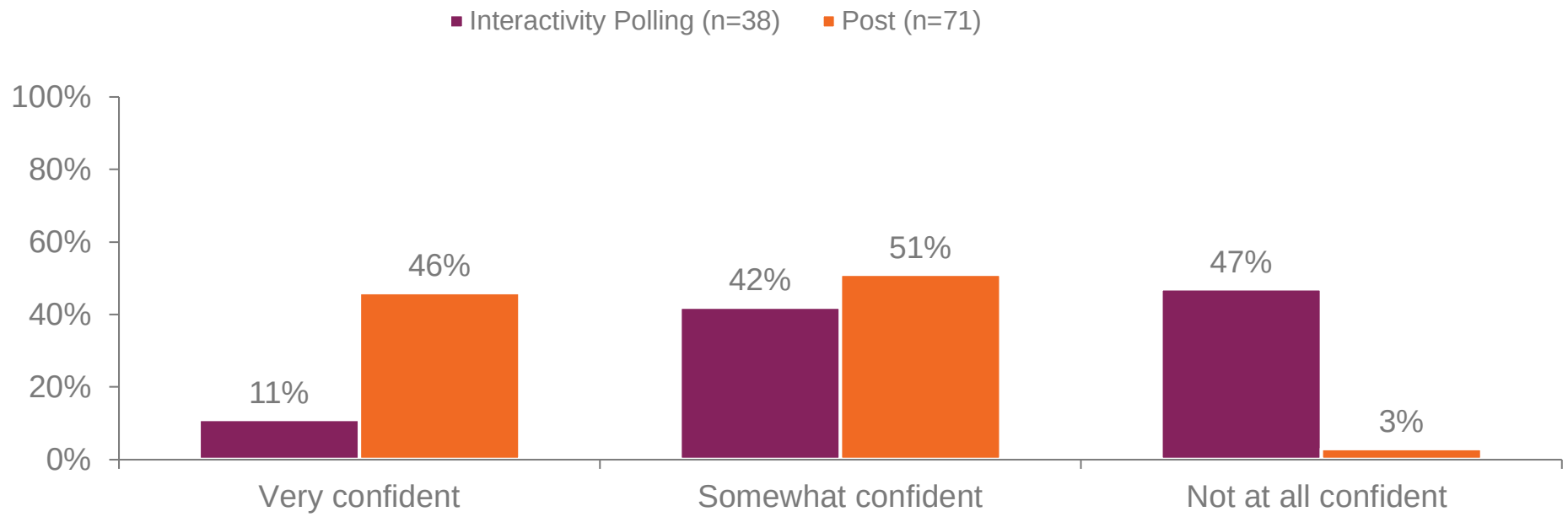
Confidence Assessment: CF-ILD Tx

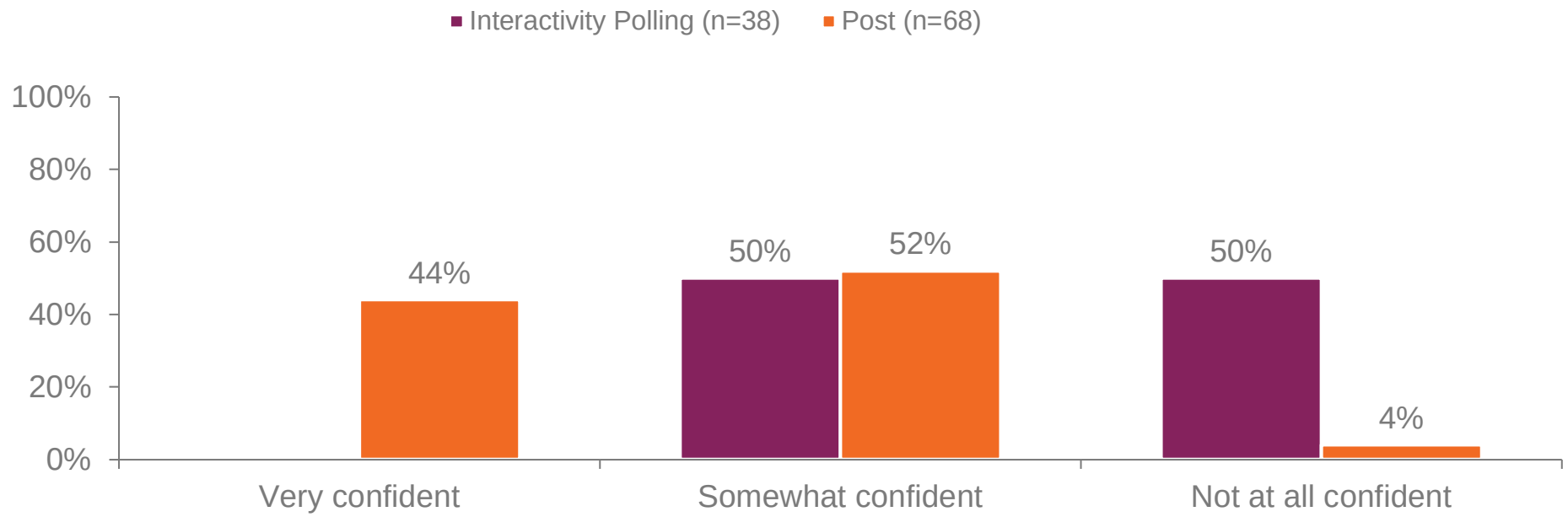
How confident are you in discussing available therapy and supportive care with patients with CF-ILD?



There was a 318% increase post-activity in learners who self-identified as "very confident" with discussing available therapy and supportive care of patients with CF-ILD. Potential topics to be discussed with patients include FDA-approved treatment for CF-ILD, support groups, high protein after workouts, and comprehensive CF-ILD management.

Confidence Assessment: CF-ILD Mgmt

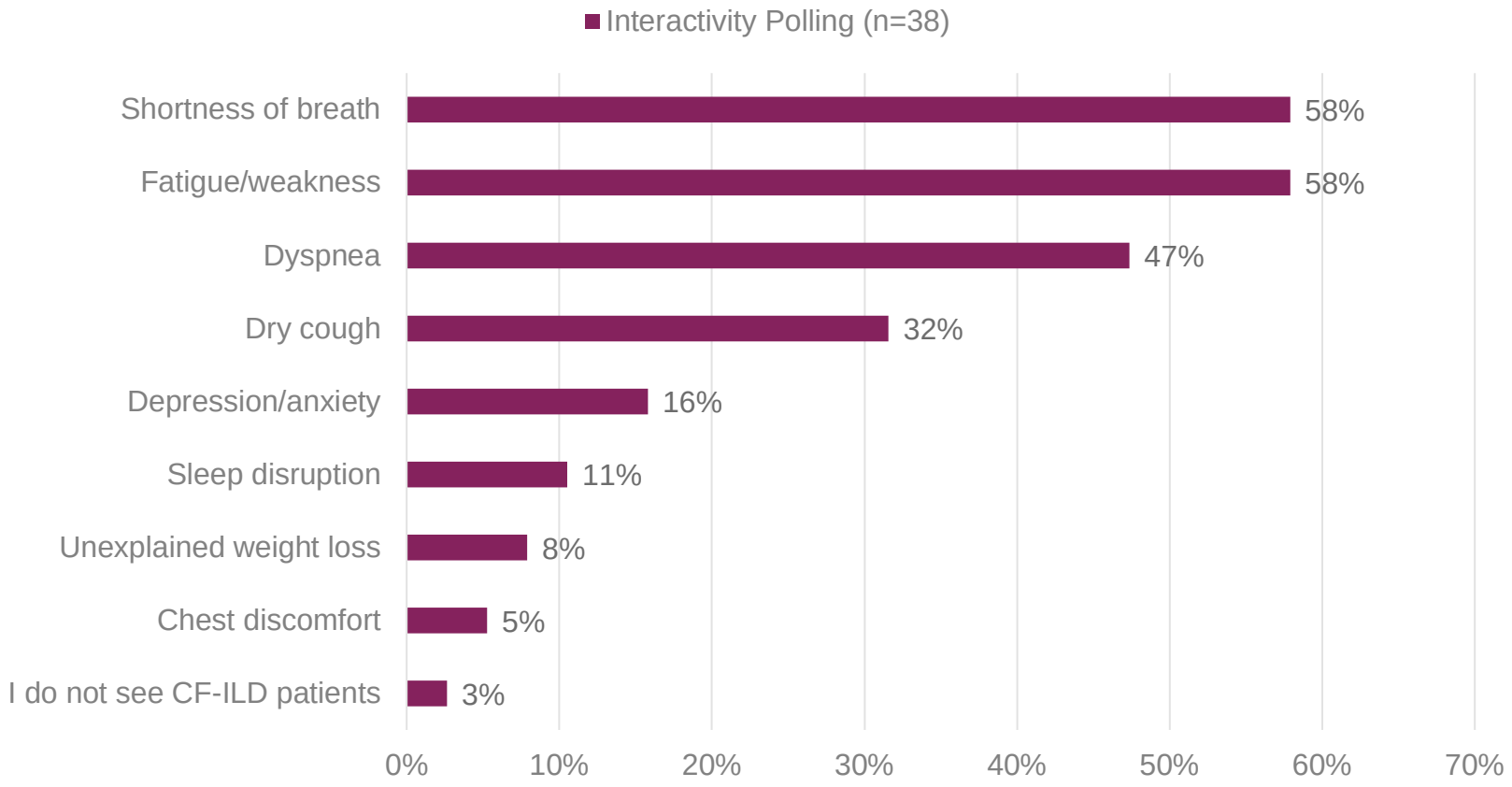
How confident are you in tailoring personalized management plans for patients with CF-ILD?



No learners (0%) pre-activity self-identified as "very confident" with tailoring personalized management plans for patients with CF-ILD; however, this number jumped to 44% post-activity. It is critical for respiratory therapists as part of the interdisciplinary care team to be familiar with the treatment plans of patients with CF-ILD to help ensure that their patients are receiving the best care and are adherent to their regimen.

Patient Symptom Assessment

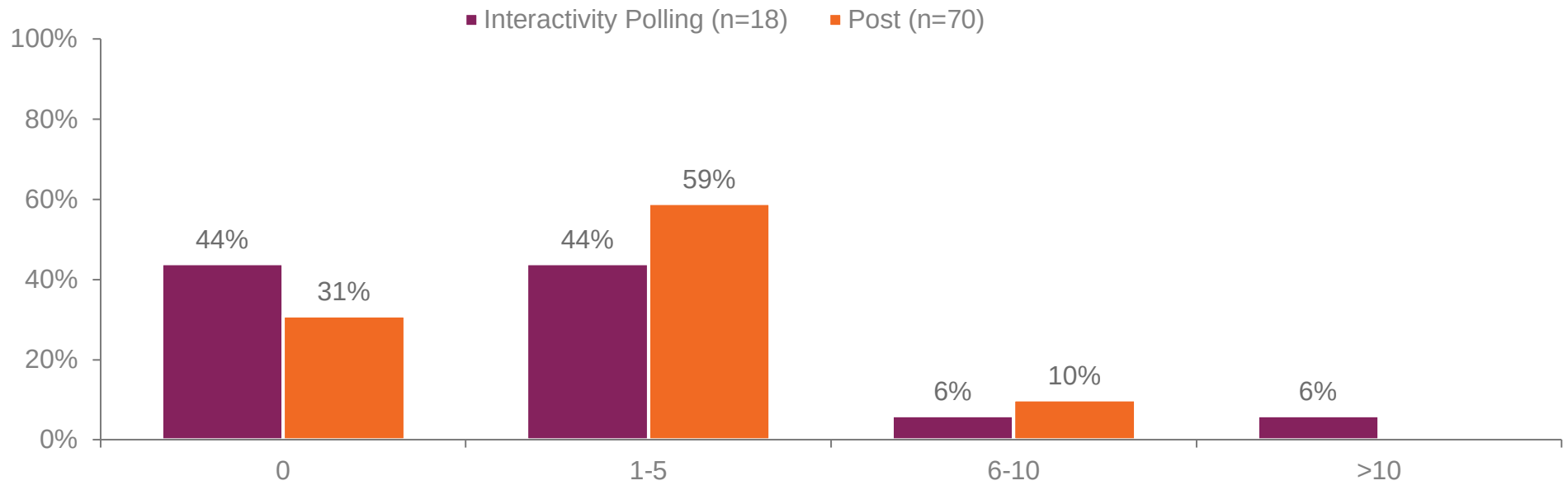
Please select the 3 most common symptoms your patients with CF-ILDs report as negatively impacting their quality of life:



Learners reported that the 3 most troublesome symptoms of CF-ILD that impact patient quality of life are shortness of breath, fatigue/weakness, and dyspnea. Dr. Cosgrove mentioned that these are the symptoms he would expect as well. Knowledge of these common impactful symptoms can inform patient-centered prioritization of comprehensive care, including pulmonary rehabilitation.

Clinical Assessments: Respiratory Disease Misdiagnoses

How many patients have you seen over the last 12 months who you suspect may have been misdiagnosed or not yet properly diagnosed with CF-ILD?



Nearly half of learners during the activity and almost two-third of learners post-activity considered 1-5 patients seen in the past year who may been misdiagnosed or not yet properly diagnosed with CF-ILD. Post-activity, 1 in 10 learners said this was the case for about 6-10 patients. These data are alarming given the morbidity and mortality rates associated with CF-ILD and the need for appropriate, timely care. This activity should motivate learners to look out for patients who may not yet be diagnosed with CF-ILD, as it is a disease more common than they may realize.

Knowledge Acquisition: CF-ILD Clinical Characteristics

CF-ILD is associated with which of the following?

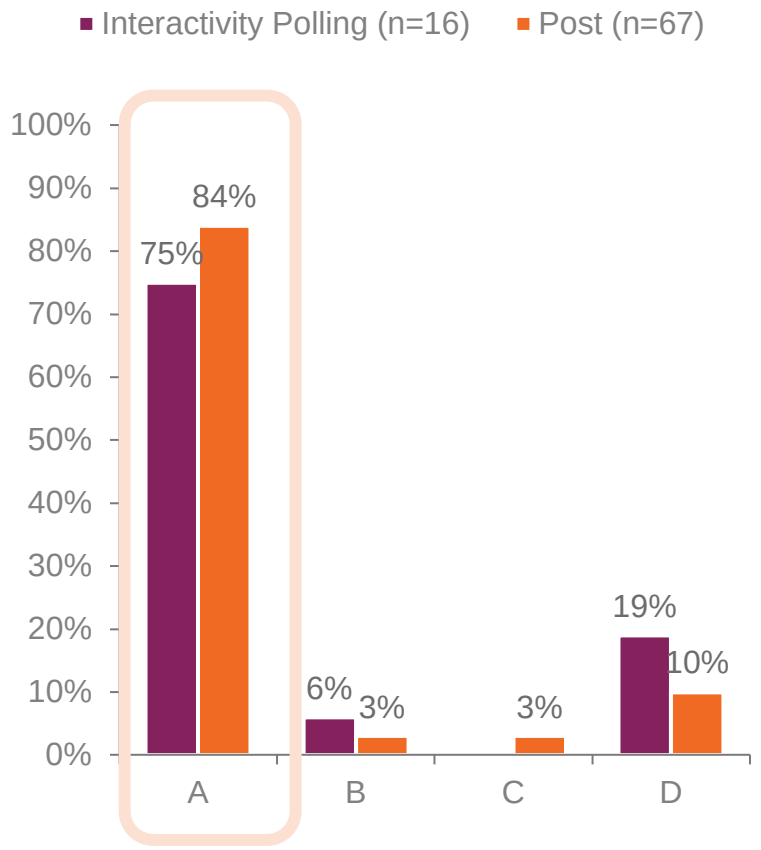
A. Decline in FVC and/or Dlco

B. Increase in FVC and/or Dlco

C. Decline in FVC only

D. Decline in Dlco only

While many learners were already familiar with the clinical features of CF-ILD, there were still increases in knowledge in this topic post-activity. It is critical for respiratory therapists to understand the characteristics of this disease to help with these patients' diagnosis and assessment and differentiation from other pulmonary diseases they may see in their clinical setting.



Knowledge & Competency Gains

Pretest vs Posttest Comparison

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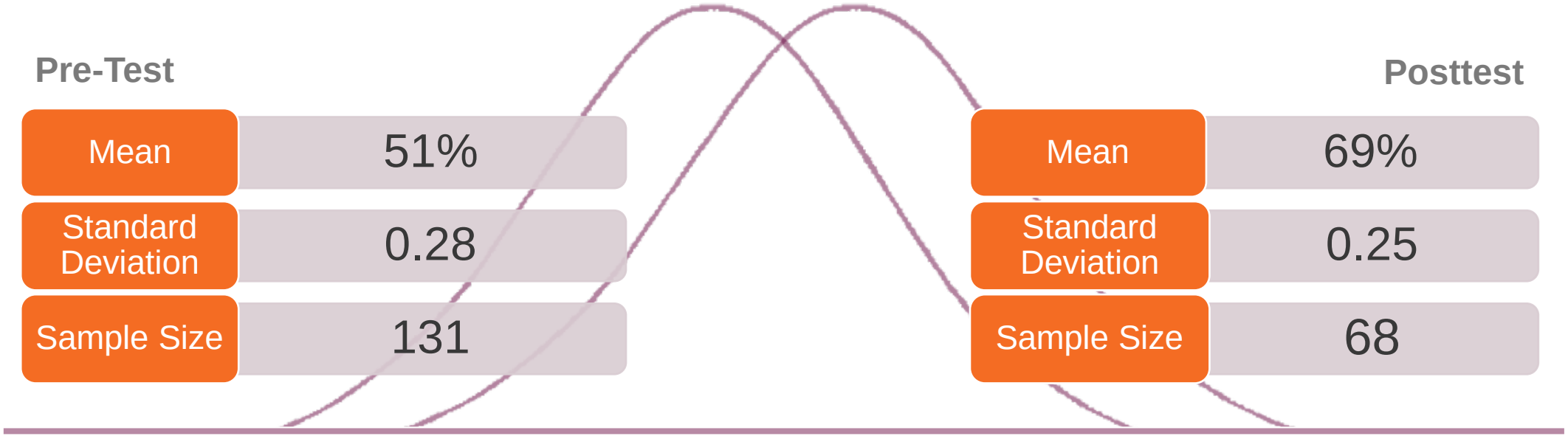
Cohen's *d* Effect Size

This Effect Size calculation uses pre/post-test question responses from all paired pre and posttest takers.



Cohen's d Effect Size = 0.68

An effect size of 0.68 indicates that participating HCPs are now ~42.04% more knowledgeable of the content assessed than prior to participating in this virtual symposium.

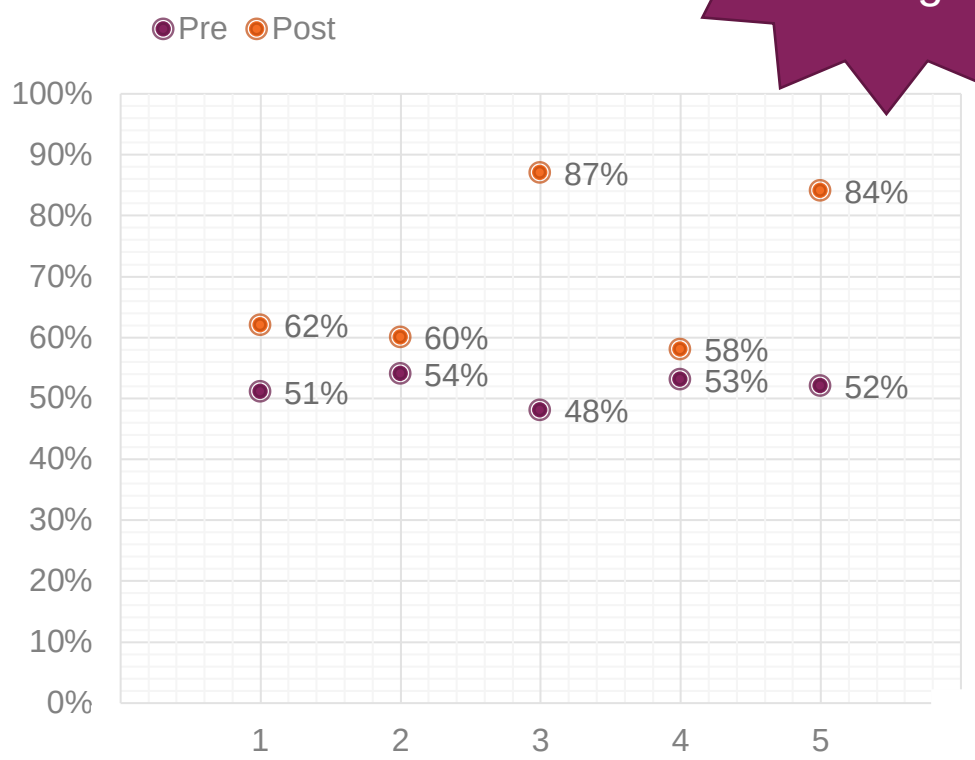
Cohen (1988): .2 = small, .5 = medium, .8 = large
Wolf (1986): .25 = educationally significant, .50 = clinically significant

Pretest vs. Posttest Summary

37% Average Change

Overview of Correct Responses

Topic	% Change*
Early CF-ILD signs & symptoms	↑ 22%
Various CF-ILD subtypes	↑ 11%
Approved CF-ILD agents	↑ 81%
Impacts of approved CF-ILD therapy	↑ 9%
Comprehensive CF-ILD care	↑ 62%



Participants demonstrated improved knowledge and competence on five of five pre/posttest questions.

*Relative percent change between pre-assessment score and post-assessment score.

Clinical Competence: Early CF-ILD Signs & Symptoms

Objective: Describe the identification of progressive-fibrosing interstitial lung disease (CF-ILD) across disease entities

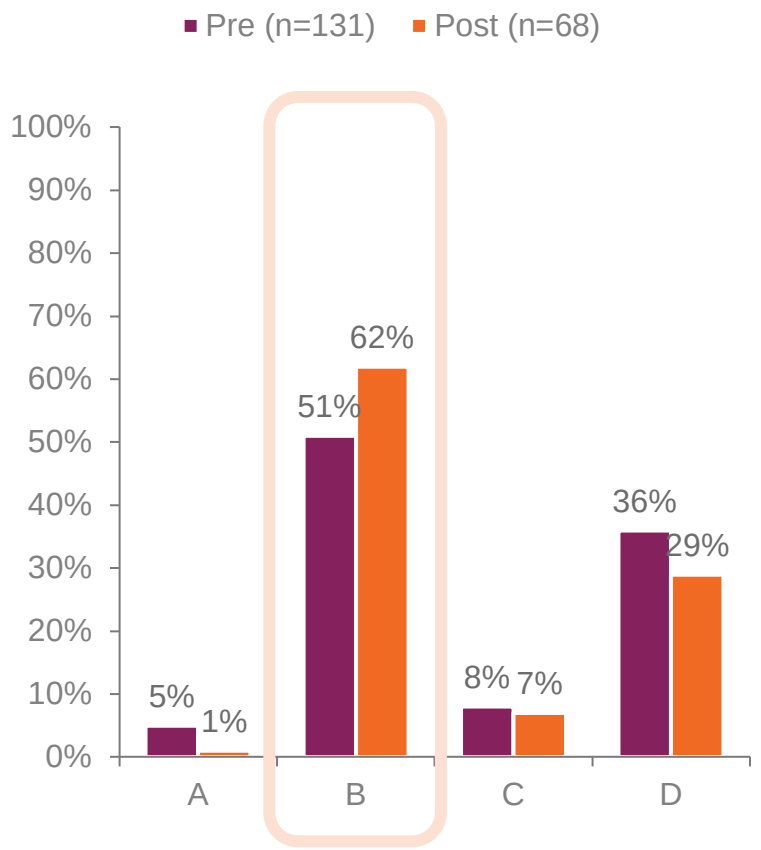
1. You are currently assessing a 61-year-old patient, Claudia with pulmonary function testing. Her doctor has diagnosed her with COPD, but you question this secondary to her symptom presentation. Her present symptoms include dyspnea; dry, hacking cough; fatigue; and weight loss. Which of the following symptoms is NOT associated with COPD and more so associated with a sign/symptom of CF-ILD?

   A. Dyspnea

   B. Dry, hacking cough

   C. Fatigue

   D. Unexplained weight loss



While about half of learners were already familiar with the early features of CF-ILD, increased competency was demonstrated in this topic post-activity. It is essential for respiratory therapists to understand the early symptoms of CF-ILD opposed to other respiratory conditions, particularly COPD, which they see frequently. Education should continue to highlight these differences.

Knowledge Acquisition: Various CF-ILD Subtypes

Objective: Describe the identification of progressive-fibrosing interstitial lung disease (CF-ILD) across disease entities

2. Which of the following conditions may progress and become associated with a CF-ILD?

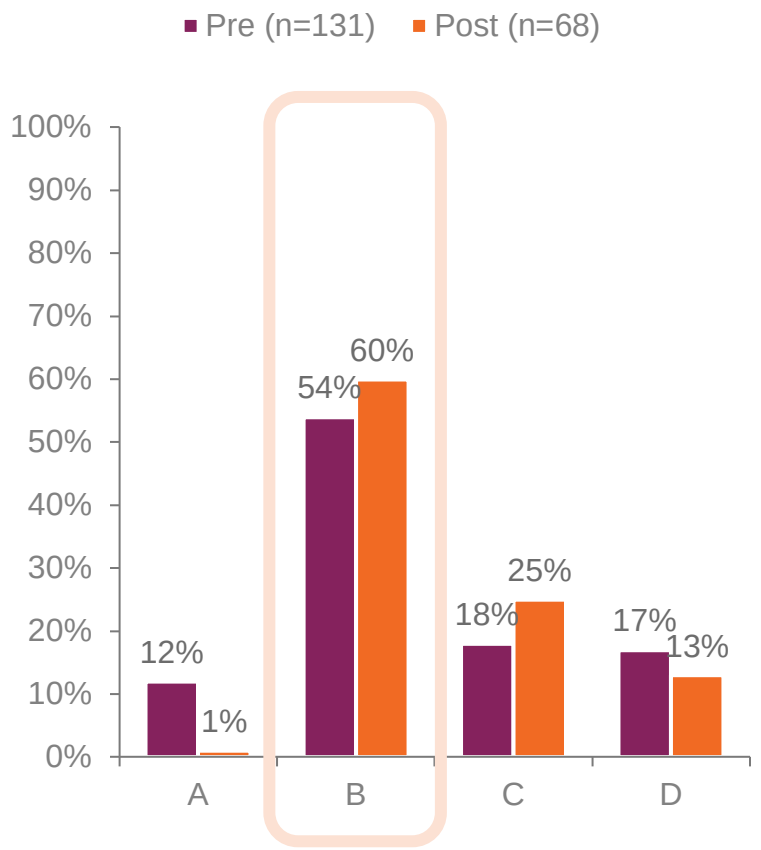
   A. Psoriasis

   B. Systemic sclerosis

   C. Sjögren's syndrome

   D. Celiac disease

Over half of learners were familiar with the different subtypes of CF-ILD at baseline and which conditions could become associated with progressive fibrosis, there was a modest increase in knowledge in this topic post-activity. Respiratory therapists need to know the conditions that are associated with CF-ILD to recognize at-risk patients and aid the interdisciplinary care team with proper diagnosis, assessment, and management.



Knowledge Acquisition: Approved CF-ILD Agents

Objective: Outline pharmacologic and respiratory therapy interventions that can assist patients with CF-ILD

3. Which of the following is the first and only approved agent for CF-ILD with a progressive phenotype?

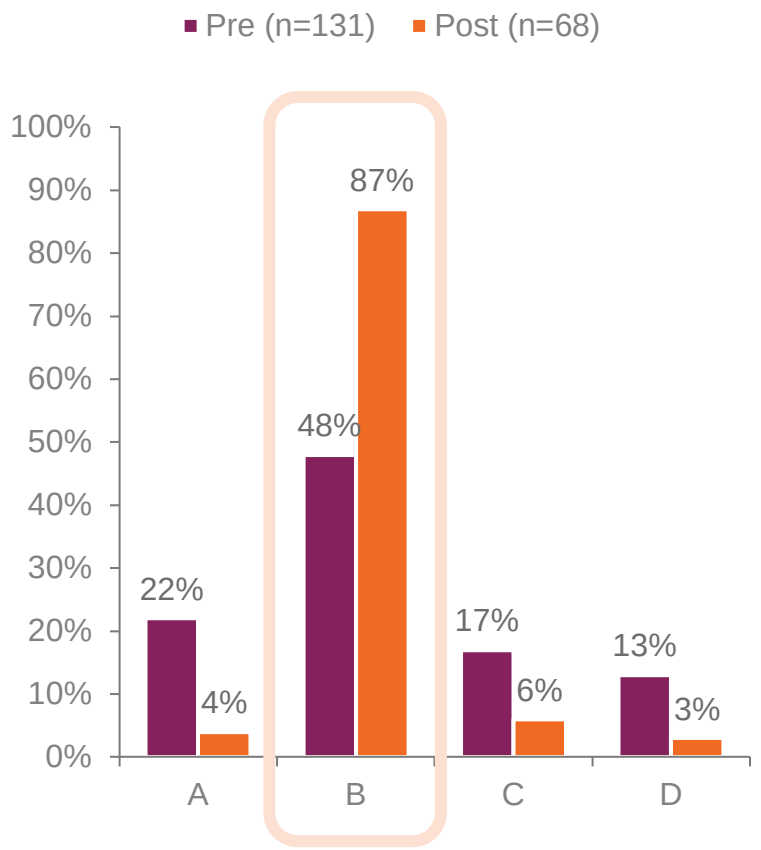
   A. Pirfenidone

   B. Nintedanib

   C. Tocilizumab

   D. Methotrexate

Learners demonstrated marked knowledge gains related to the first and only approved therapy for CF-ILD, nintedanib. It's beneficial for respiratory therapists to know which agents are available and effective for their patients with CF-ILD to ensure that their patients are receiving the most optimal care.



Knowledge Acquisition: Impacts of Approved CF-ILD Therapy

Objective: Outline pharmacologic and respiratory therapy interventions that can assist patients with CF-ILD

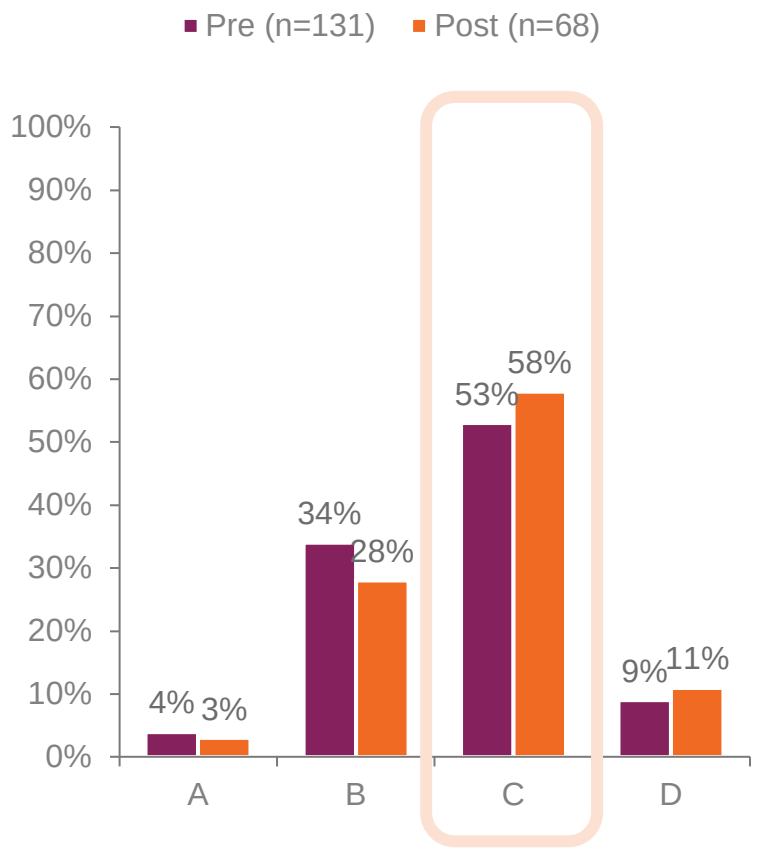
4. A patient who is considering treatment with the FDA-approved antifibrotic therapy for CF-ILD should be counseled that:

   A. It will make them feel better

   B. It may cause side effects that can be managed, but it will decrease the risk of mortality

   C. It delays disease progression, but they may not notice a difference in symptoms

   D. It will reduce some of their symptoms



While about half of learners were already familiar with the impacts of approved CF-ILD therapy, there were still increases in knowledge in this topic post-activity. Respiratory therapists must understand the efficacy of the approved CF-ILD agents to help patients understand treatment goals and expectations.

Knowledge Acquisition: Comprehensive CF-ILD Care

Objective: Discuss the impact of CF-ILD on patient quality of life

5. It is important for patients with CF-ILD to consume which of the following after exercise?

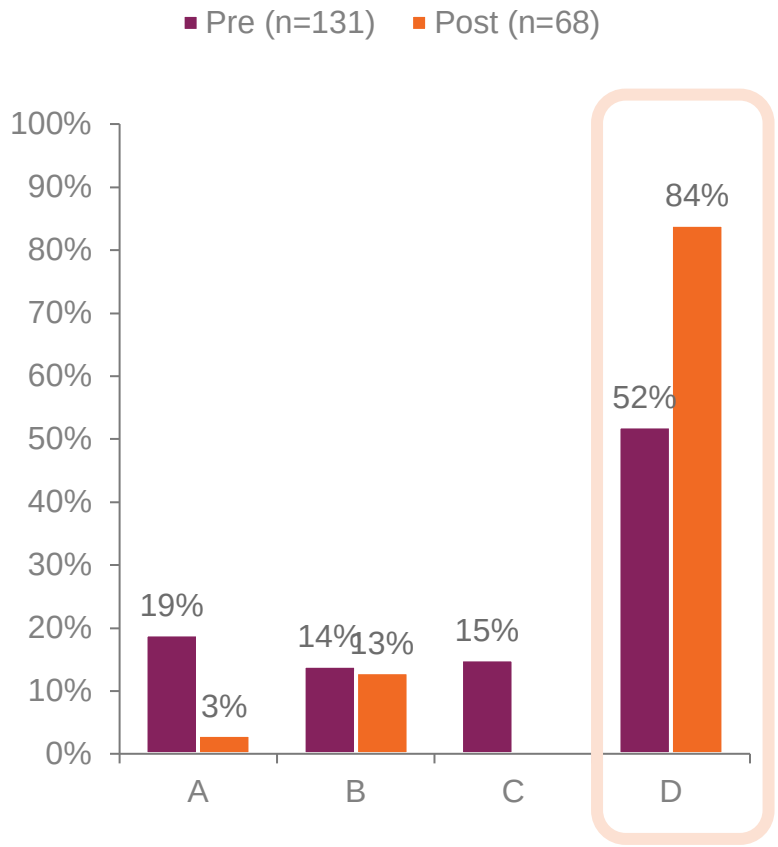
   A. Carbohydrates

   B. Fats

   C. High calorie snacks

   D. Protein

Learners experienced knowledge gains related to comprehensive CF-ILD care that can potentially improve quality of life, specifically strategies that can support effective pulmonary rehabilitation. Respiratory therapists must also understand that they need to treat their patients holistically as quality of life is impacted by more than just their lung health.



Practice Change, Patient Impact and Learner Engagement

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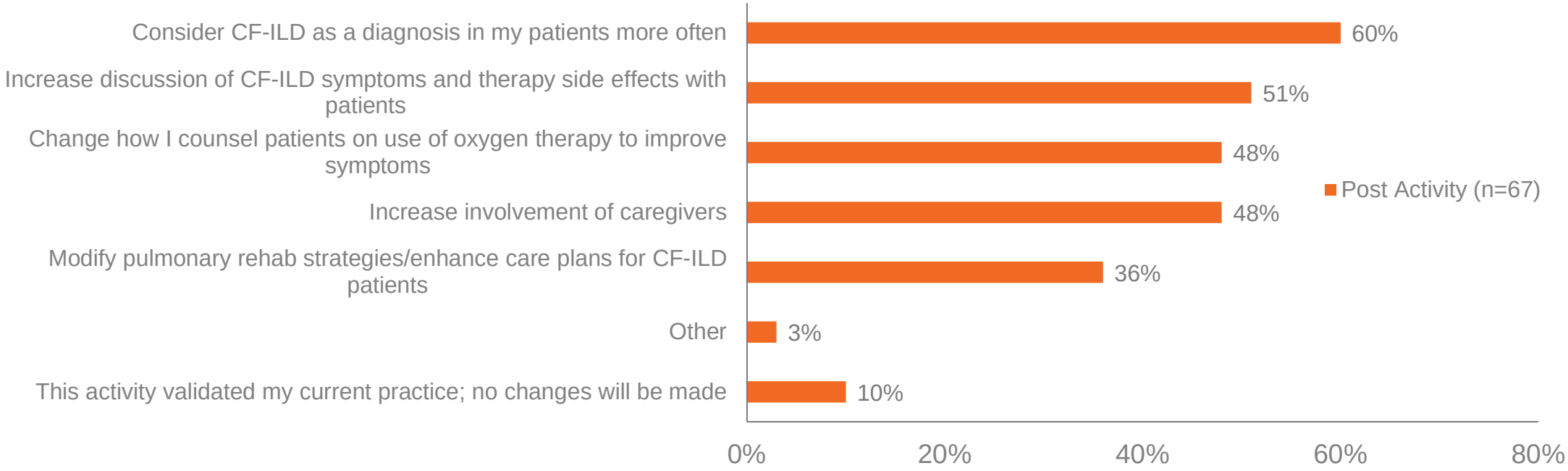
Self-reported Impact

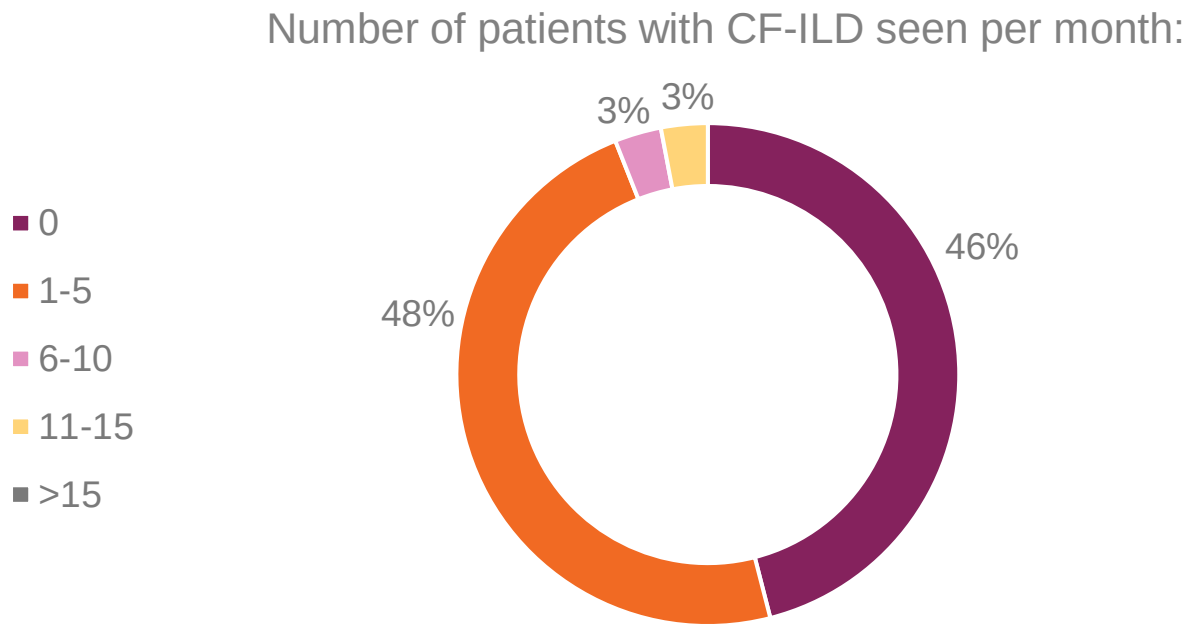
Practice Change



90% of learners committed to making changes in their practice immediately following their participation. Specifically, 60% will more often consider CF-ILD as a diagnosis in their patients and 51% will increase discussion of CF-ILD symptoms and therapy side effects with their patients.

Multiple responses allowed

Patient Care Impact

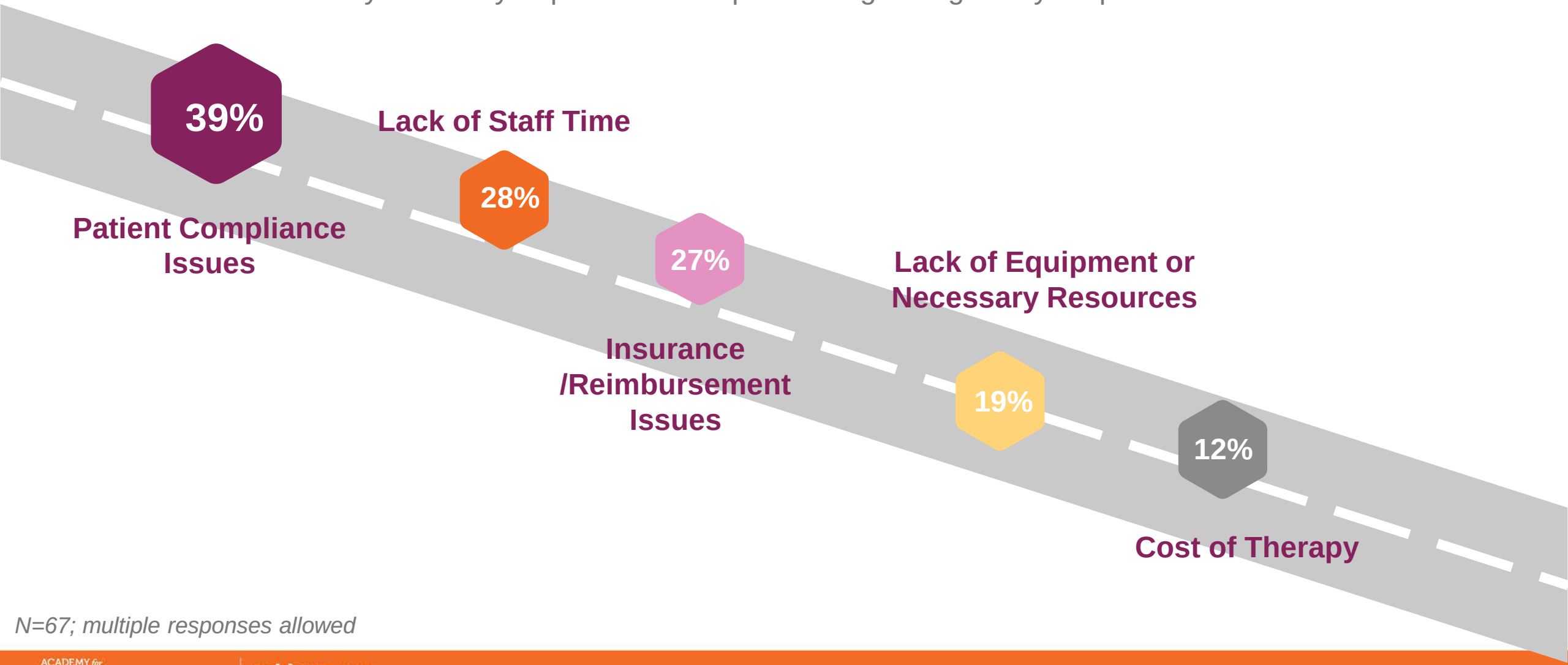


Changes will impact 125 to more than 397 patients with CF-ILD each month. This assumes data in chart above is representative of all symposium participants (140), who indicated they would change their practice as a result of their participation in this activity (90%).

N=67

Self-reported Barriers to Change

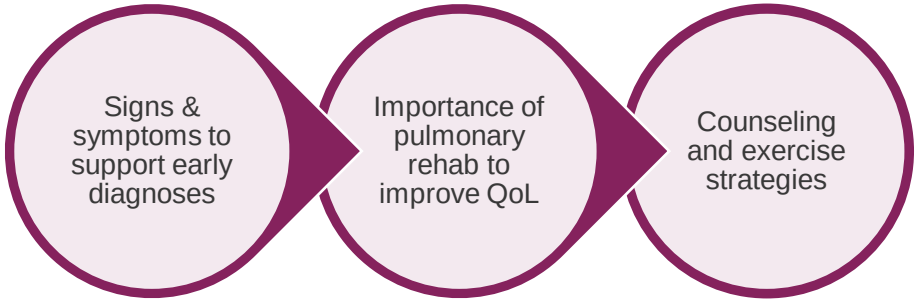
Please indicate any barriers you perceive in implementing changes in your practice:



N=67; multiple responses allowed

Activity Impact

What is one pearl you took away as a result of your participation?



- All patients are different, and treatment must be tailored to the individual and his/her disease process
- Auto-immune plays a major roll in some lung issues.
- Better understanding of how to treat ILD
- General deepening of my understanding of ILD
- High protein after workout
- How important pulmonary rehab is to ILD patients
- How important rehab is for this diagnosis in improving pt. QOL.
- I realize that I may want to be part of a pulmonary rehab as a change in my focus on my career.
- Importance of support groups
- Improving a patient's quality of life and outlook on life
- Improving quality of life is paramount
- It is more prevalent than most of us realize
- It was the way in which the speaker related information to the patient. I practiced saying it a few times!
- Learning more about how CF-ILD patients deal with their disease
- Patient prospective
- Pulmonary Rehab and exercise is important to improving quality of life
- Role of the resp. therapist in pulmonary rehabilitation.
- Support Group participation/referral importance
- That DME companies are required to fill the prescription of oxygen to its full extent
- That there are many aspects of this disease and too many times they are not considered.
- The difference between COPD and CF-ILD
- The importance of pulmonary rehab and use of the newest med.
- There is a medication that is specifically targeted for specific CF-ILD patients
- There was a nice light shed on how diagnosing this is a process and may change as more symptoms arise.
- This session brought increased consideration of CF-ILD as a potential diagnosis in our patient population.
- Treat patient not disease
- Understanding that it is a "marathon" not a sprint to explaining the treatment plan to the patients.
- Understood better on how to counsel a CF-ILD patient
- Use of the St George Questionnaire, another tool to give students.
- Valuable information so that diagnosis isn't missed in the future.

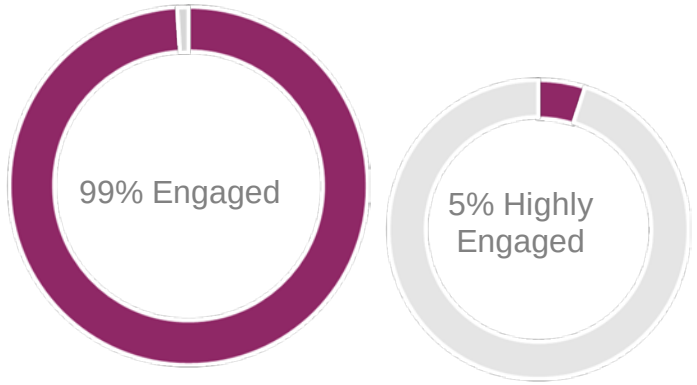
Activity Impact

What was your main motivation for participating in this activity? And was it met?

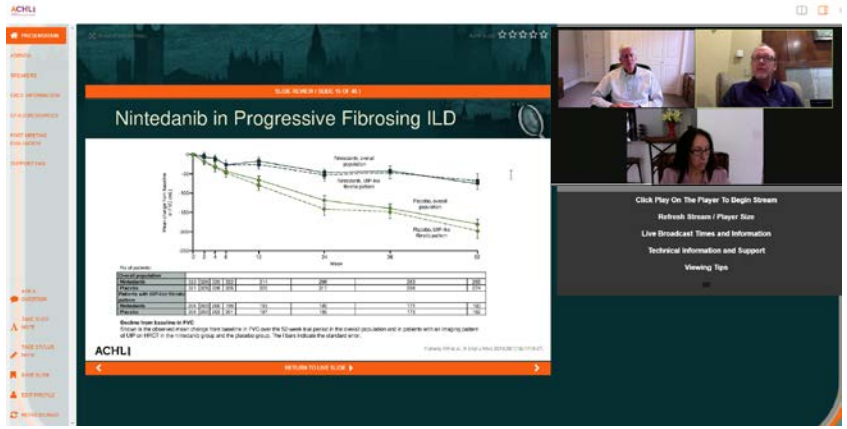
- I have a specific patient - answered questions about new therapy
- AAC credits from a course I would learn from- Yes. I enjoyed the program and learned new things about CF-ILD and can help my patients with the info.
- ***As an RT in a Pulmonologist office, I'm involved in patient education, testing and setting up DME. These all helped that I am not alone in the roadblocks for best care for theses patients. Yes, it was met and exceeded.***
- Better knowledge on Management of CF-ILD patient. Yes
- CEU for recertification as well as neuroplasticity
- Continuing Education
- Education about CF-ILD
- Education and yes it was very informative.
- education, yes
- Expanding my understanding in this field of respiratory therapy.
- find out more information, yes
- Gain a better understand in helping these people beyond the office.
- Gain knowledge and confidence- Yes
- Gaining more information to teach students, and yes it was met.
- Get knowledge CF ILD
- ***I had one patient I would have put into this category and I had one year to learn from our experiences before he died. His DLCO was at 29% 5 years prior to meeting me and his primary care doctor did not have him follow up with a pulmonary physician.***
- I had very little understanding of the diversity that surrounds our ILD patients.
- I wanted to see if there were new revelations in the diagnosis and management of this disease process. Yes
- Increase knowledge and yes
- Increase knowledge of ILD and it was.
- Increase my knowledge yes
- Increasing knowledge
- Interest over the years in this disease, an opportunity to see how it is being treated currently and the opportunity to obtain CEU credit.
- Knowledge of CF ILD- was met Thank you to three speakers- wonderful session
- Learn more about treatment/management. Yes
- Learning
- learning more about how CF-ILD patients deal with their disease
- ***Lessen my knowledge gap about CF-ILD. I am pleased to have this updated information to use with my patients.***
- My dad died of IPF. Wanted to hear the new info. Yes.
- Pulmonary rehab strategy, yes
- to better informed to assist patients in the future.
- To gain knowledge
- To hear how the respiratory therapist deals with pulmonary rehab. Yes
- To learn more about CF-ILD, yes
- To understand interstitial lung disease better. Yes
- Wanted to know more about PF and treatment. Great presentation.
- Wanting to understand ILD
- Yes, the recent encounters of Sjogrens, Rheumatoid, Lupus patients seen, this past year.

Learner Engagement & Insights

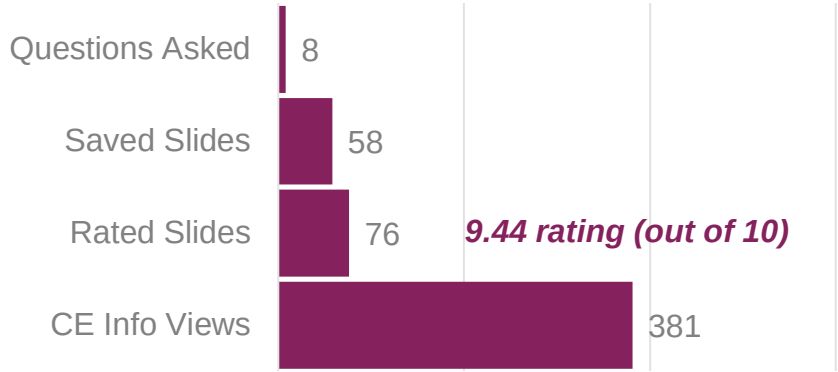
This virtual symposium incorporated the Array™ learning system to engage learners to follow along, take notes, save slides, and ask questions. Upon signing in, all learners were able to view the custom learning platform on their own desktops and mobile devices.



*Of the 140 participants, 99% (139) were **Engaged** with the ARRAY™ technology, meaning these participants viewed and saved content, responded to polling, etc. Those who were **Highly Engaged** took it to the next level of engagement such as asking questions and taking notes, which accounted for 5% (7) of learners.*



Understanding how participants engaged with the presentation provides insight to improve and tailor future education.



N=140

Top Rated Slides

Top 3 Saved Slides

Home Exercises During COVID-19 Pandemic

- Resphealth.org/pulmonary-rehabilitation-at-home/
- Nyp.org/rehabmed/rehab-exercise-videos
  - See: "Pulmonary Rehab Home Program" video
- Cpff.ca/covid-19/videos/
- Pulmonarywellnessonline.com

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Exercise

- Aerobic exercise
- Breathing retraining
- Upper and lower extremity exercises
- Strengthening exercises
- Endurance training
- Oxygen titration during exercise for ILD patients
- Importance of protein after exercise

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Education

- Signs and symptoms of exacerbations
- Nutrition
- Exercise
- Emotional control (anxiety and depression)
- Lung
- Breathing retraining (PLB and diaphragmatic technique)
- Lung disease
- Oxygen therapy

PLB = pursed-lip breathing

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Top 3 Slides with Most Notes

Risk Factors

RA-ILD	Primarily male
SSc-ILD	Usually have diffuse cutaneous SSc, African-American ethnicity, older age at disease onset, shorter disease duration, presence of anti-Scl-70/anti-topoisomerase I antibody and/or absence of anticentromere antibody
HP	More common in women and those older than 65 years old
IPF	Primarily male
Idiopathic non-specific interstitial pneumonia	Usually present in mid-adulthood, are female, and more than half have never smoked
Sarcoidosis	Predominantly in 30-40-year-olds, more commonly aggressive in patients of Afro-Caribbean descent than in Caucasians
Myositis-related ILD	30-40% of patients with myositis have some form of lung disease, with ILD being the most common and serious complication of inflammatory muscle disease
Sjogren's syndrome-related ILD	More likely to be male, older, and smokers
Unclassifiable ILD	Mean age of 68 years, 64% former smokers

RA = rheumatoid arthritis; SSc = systemic sclerosis

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Expert Guidance Summary for CF-ILD Treatment

- Choice of therapy often depends on patient tolerance
- Preferred agents are determined by underlying cause
- Current data suggest that all patients have some level of benefit from antifibrotics, but immunosuppressants require treatment trials to assess risk (side effect)/benefit
- Treatment decisions are always a "shared decision-making process" involving risk vs. benefit
  - Discussion starts after diagnosis
  - If lung function is good, can wait and monitor as many patients (eg, with RA) may have no progression at all
- Serial monitoring every 3-6 months with PFTs

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Support for Pulmonary Rehabilitation

Study	Endpoints
Swigris J – 6-8 weeks; 60% max *Eur Resp J* 2011	↑ Functional capacity and decreased fatigue
Nishiyama O – 10 weeks; 80% max *Respirology* 2008	6MWT ↑ 46 M
Vainshelboim B –12 weeks *Arch Phy Med Rehabil* 2016	Exercise training improves exercise tolerance, functional capacity, pulmonary function, dyspnea and QoL in patients with IPF, suggesting a short-term treatment efficacy for clinical improvement, and should be considered the standard care for IPF.
Huppman P – ILD *Eur Respir J* 2013	6MWT ↑ 46 M; no change in dyspnea ratings, improved QoL
Holland AE – ILD and IPF *Cochrane Review* 2014	6MWT ↑ 44 M on average Maximum exercise capacity, shortness of breath and, QoL

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Top Rated Slides (cont)

Top 3 Slides with Most Questions

Patient Interview

Could you please describe your diagnostic journey? Did you receive other diagnoses before CF-ILD? If you did, what were those misdiagnoses?

What were some of the biggest questions and concerns you had when you were diagnosed?

What has your treatment experience been like? What drugs have you been treated with, and what are you being treated with now? What has your progress been like with these agents?

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Morbidity and Mortality

RA-ILD	Median survival = 5 years from time of initial clinic visit
SSc-ILD	Median survival = 11.2 years from the date of first HRCT showing evidence of ILD
HP	Only 58% of patients alive ~7 years after diagnosis
IPF	Mean post-diagnosis survival = 3-4 years
Idiopathic non-specific interstitial pneumonia	2-, 5-, and 10-year survival is 87.7%, 60.7%, 35.9%, respectively
Sarcoidosis	5- and 10-year survival is 3.9% and 9.0%
Myositis-related ILD	1-, 5-, and 10-year survival is 97%, 91%, and 81%, respectively
Sjogren's syndrome-related ILD	NSIP is most common subtype (45%). 5-year survival is 83%
Unclassifiable ILD	1-, 2-, and 5-year mortality rates are 10.6%, 23.8%, and 31.1%, respectively
Coal worker's pneumoconiosis	Years of potential life lost of life expectancy per decedent = 12.6 years

HRCT = high-resolution computed tomography

Kolb M et al. *Respir Res*. 2019;20:57; Nunes H et al. *Eur Respir J*. 2018;48:746-758; Kinkl G et al. *Chest*. 2018;153(1):105-113; Johnson C et al. *Lung*. 2016;194(6):733-7; Flament T et al. *Eur Respir Rev*. 2018;28:110-122; Mazurek JM et al. *MMWR Morb Mortal Wkly Rep*. 2018;67:819-824; Ryerson CJ et al. *Eur Respir J*. 2012;40:750-787.

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Patient Interview

What are some challenges that you experience with daily activities of living?

How do you cope with these daily living challenges? (eg, changes you have made, techniques, strategies you have learned)

What are some other barriers or challenges that you continue to experience? (These can also be related to your patient care or even to the COVID-19 pandemic)

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Top 3 Most Engaging Slides

Home Exercises During COVID-19 Pandemic

- Resphealth.org/pulmonary-rehabilitation-at-home/
- Nyp.org/rehabmed/rehab-exercise-videos
  - See: "Pulmonary Rehab Home Program" video
- Cpff.ca/covid-19/videos/
- Pulmonarywellnessonline.com

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Exercise

- Aerobic exercise
- Breathing retraining
- Upper and lower extremity exercises
- Strengthening exercises
- Endurance training
- Oxygen titration during exercise for ILD patients
- Importance of protein after exercise

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https://www.lung.org/lung-health-diseases/lung-disease-lookup/pulmonary-fibrosis/patients/living-well-with-pulmonary-fibrosis/physical-activity. Accessed October 14, 2020.

Essential Components of Pulmonary Rehabilitation

- Assessment
- Supervised therapeutic exercise
- Education
- Psychosocial intervention
- Long-term adherence

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https://www.lung.org/blog/pulmonary-rehab-helps-breathe. Accessed October 14, 2020.

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