

### **Did the authors miss any major topic(s)?**

The role of the Physical Therapist (PT) in caring of pwCF has been overlooked in this paper. I would recommend that PT be moved to the Core Team. Based on the goals and aims of this paper, PTs are essential to providing care that addresses the whole person, supporting participation, quality of life and improved outcomes.

Exercise and physical activity are essential to a healthy physical, mental and emotional life! All human beings must move in order to participate in life, from the youngest infant who must learn to hold a bottle or roll over, to the oldest person who gains joy from gardening, to the person who dreams of climbing mountain peaks. Physical Therapists are experts in helping people reach these goals through individualized assessment, prescription, monitoring and adapting exercise, considering the physical, mental and social situations unique to each individual. As a core member of the CF Care Team, the PT can integrate the laboratory test results, imaging results, lung function, nutritional function, and mental/emotional health status along with the results of exercise assessment to develop a safe, effective plan of movement that helps each person with CF meet their individual goals. In order to effectively gather and integrate this information, it is critical that the PT be an active member of the interdisciplinary team communication that happens in the CF clinic (and via telehealth), to integrate their findings and recommendations in real-time with patients and families.

The CF Care Team PT also relies on a referral network of PT specialists, much in the way that the CF physician relies on their colleagues in other areas of medicine. For example, the CF Care Team PT might refer to a Sports Clinical Specialist for training a patient for athletic participation, or to a Pelvic Health Clinical Specialist for a pregnant patient. PT Clinical Specialists in Pediatric, Geriatric, Orthopedic, Neurologic, Oncologic, Clinical Electrotherapeutics/Wound Care, and Cardiovascular & Pulmonary specialty practice areas may be consulted for patients with development/aging issues, musculoskeletal or neurologic conditions, wound healing, vestibular rehab, critical illness or cancer complications to their CF disease. Thus, the CF Care Team PT has teams of experts at their disposal, although the Care Team PT is the expert in the effect of CF on the components of movement for each individual patient.

### **Did the authors miss any major citation(s)?**

1. Button BM, Wilson C, Dentice R, Cox NS, Middleton A, Tannenbaum E, Bishop J, Cobb R, Burton K, Wood M, Moran F, Black R, Bowen S, Day R, Depiazzi J, Doiron K, Doumit M, Dwyer T, Elliot A, Fuller L, Hall K, Hutchins M, Kerr M, Lee AL, Mans C, O'Connor L, Steward R, Potter A, Rasekaba T, Scoones R, Tarrant B, Ward N, West S, White D, Wilson L, Wood J, Holland AE. Physiotherapy for cystic fibrosis in Australia and New Zealand: A clinical practice guideline. *Respirology* 2016; 21: 656-667.

2. Gruet M, Saynor Z, Urquhart D, Radtke T. Rethinking physical exercise training in the modern era of cystic fibrosis: A step towards optimising short-term efficacy and long-term engagement. *J Cyst Fibros*. 2022; 21(2):e83-e98.
3. Saynor Z, Gruet M, McNarry M, et al. Guidance and standard operating procedures for functional exercise testing in cystic fibrosis. *Eur Respir Rev*. 2023 Aug 9;32(169):230029
4. Cox NS, Alison JA, Button BM, Wilson JW, Morton JM, Holland AE. Accumulating physical activity in at least 10-minute bouts predicts better lung function after 3-years in adults with cystic fibrosis. *ERJ Open Res* 2018; 4.
5. Gruet M, Troosters T, Verges S. Peripheral muscle abnormalities in cystic fibrosis: Etiology, clinical implications and response to therapeutic interventions. *J Cyst Fibros* 2017; 16: 538-552.
6. Nixon PA, Orenstein DM, Kelsey SF, Doershuk CF. The prognostic value of exercise testing in patients with cystic fibrosis. *N Engl J Med* 1992; 327: 1785-1788.
7. Ionescu AA, Evans WD, Pettit RJ, Nixon LS, Stone MD, Shale DJ. Hidden depletion of fat-free mass and bone mineral density in adults with cystic fibrosis. *Chest* 2003; 124: 2220-2228.
8. Prevotat A, Godin J, Bernard H, Perez T, Le Rouzic O, Wallaert B. Improvement in body composition following a supervised exercise-training program of adult patients with cystic fibrosis. *Respir Med Res* 2019; 75: 5-9.
9. Gruber W, Stehling F, Olivier M, Dillenhofer S, Koerner-Rettberg C, Sutharsan S, Taube C, Mellies U, Welsner M. Effects of a long-term exercise program on motor performance in children and adolescents with CF. *Pediatr Pulmonol* 2020; 55: 3371-3380.
10. Fernanda, M et al. Peak oxygen Uptake and Mortality in Cystic Fibrosis: Systematic Review and Meta-Analysis. *Resp Care*. 2019. Vol 64, 91-99.
11. Hebestreit H, Arets H, Aurora P, Boas S, Cerny F, Hulzebos E, Karila C, Lands L, Lowman J, Swisher A, Urquhart D. Statement on exercise testing in cystic fibrosis. *Respiration* 2015;90:332-51 DOI: 10.1159/000439057
12. Swisher AK, Hebestreit H, Mejia-Downs A, Lowman JD, Gruber W, Nippins M, Alison J, Schneiderman J. Exercise and habitual physical activity for people with cystic fibrosis: An expert consensus-based guide for advising patients. *Cardiopulm Phys Ther J* 2015;26:85-98. DOI: 10.1097/CPT.0000000000000016
13. Swisher, AK., Moffet, K. The effect of coaching on physical activity and quality of life in children and adolescents with cystic fibrosis: A quality improvement pilot study. *Internet Journal of Allied Health Sciences and Practice*. April 2010. Volume 8 Number 2.
14. Swisher AK Not just a lung disease: Peripheral muscle abnormalities in cystic fibrosis and the role of exercise to address them. *Cardiopulm Phys Ther J* 17(1):10-15, 2006.
15. Gruet M, Saynor Z, Urquhart D, Radtke T. Rethinking physical exercise training in the modern era of cystic fibrosis: A step towards optimising short-term efficacy and long-term engagement. *J Cyst Fibros*. 2022; 21(2):e83-e98.

**Please provide any feedback on the abstract and introduction: Lines 46-78**

Physical therapy should be listed here (line 68)

**Please provide any feedback on the methods: lines 79-90**

It would be good to list the CF care team member disciplines that worked on this paper (line 81).

Lines 82-84:

"A review of relevant literature was completed by each discipline with the goal of guiding potential adaptations to the CF center staffing and services."

Please be more specific with the evidence that was reviewed (categories- lung function, nutrition, exercise, bone health, GI complications) and disciplines.

**Please provide any feedback on the section: Specialized CF Care Center: Lines 91-122**

Lines 92-95:

"CF care centers offer specialized evaluation and management focused on timely diagnosis, optimizing growth and development, minimizing disease progression, and preventing/treating CF complications to allow for long and fulfilling lives. Their focus is on the individual with CF and their family/caregivers." This is a great goal/aim! However, if this is the goal of your paper, there is a lot of evidence in the literature that supports the role of PT on the care team in improving outcomes and QOL that wasn't included.

Everything in the above statement is congruent with the current practice of PTs in CF Care Centers. PTs are already key members of many care centers providing 'specialized evaluation' and interventions. Specialization is supported by: PT Mentor Program, representation on the NACFC planning committee, representation on the Education Committee, the PT grant to partially fund PTs in CF clinics, discipline-specific programming (NACFC and virtual webinars), and key documents to guide our practice (CF PT 101, Clinical Practice Guidelines).

PTs practice in interdisciplinary teams in multiple settings routinely and effectively, and are well-qualified to specialize in CF and work on CF Care teams.

Lines 119-120:

"...whereas adult programs are anticipated to grow over the next two decades, indicating an increased need for adult CF care team members."

I am in agreement with this statement and see it as a major opportunity to increase the role of PT on the care team. There are multiple unique needs of the aging CF population which PT is ideally suited to screen/treat: bone health, pelvic health and pregnancy,

menopausal changes (decline in lung function, pelvic pain, changing exercise needs), musculoskeletal conditions that could be unique to developing in the context of a pulmonary disease, vestibular hypofunction due to exposure to ototoxic drugs combined with age-related declining compensatory strategies to maintain balance, prevent falls and promote functional independence.

**Please provide any feedback on Patient-Centered Medical Homes, lines 123-136**

Lines 124-127:

"Patient-centered medical homes (PCMH) are known to improve outcomes and enhance the quality of care for individuals with chronic conditions (29154464) and multiple medical complications (32767035). These models offer comprehensive outpatient services, covering acute, chronic, and preventive care (38145573)".

This is another area that supports the inclusion of PT on the core team. PT adds comprehensive services to treat comorbidities and complications (incontinence, vestibular dysfunction, constipation, gross motor delays, pain, low bone mineral density, and impaired endurance and exercise capacity), as well as preventative care.

**Please provide any feedback on the section: Core Team, lines 137-221**

Lines 138-139:

"High-performing CF programs require capable leadership that fosters communication, collaboration, and adherence to evidence-based standards of care amongst the CF care team and pwCF"

See list of previous references providing evidence to support the role of PT for pwCF

Lines 157-159:

"Nutrition significantly impacts overall health, with a noted correlation between nutritional status and pulmonary function (17467551, 32565399)". What is the most recent evidence on this?

The references aren't listed, so I can't see the dates of the publications cited. I question if this is becoming an outdated statement in the era of modulators.

Lines 159-161:

"...there is now an increased focus on maintaining a healthy body mass index (BMI) with a rise in overweight and obesity diagnoses associated with the use of CFTR modulators.

This aligns with what PT can do! Diet alone is not enough to address overweight and obesity diagnoses, nor other conditions related to cardiovascular disease. Additionally, BMI does not distinguish between muscle, bone mass or fat. Further measures of body composition (e.g., bioelectric impedance analysis) can determine fat free mass to aid dietitians AND PTs in creating an individualized treatment plan - inclusive of diet and exercise - for optimizing body composition, rather than a focus solely on weight/BMI.

Line 164:

“CF respiratory therapists (RT), whose role is occasionally fulfilled by physical therapists...”

While PT and RT scopes of practice overlap in the area of airway clearance, they are completely different professions. No PT acts as an RT. The two professions have a different scope of education (RT primarily lungs, PT the whole body); different degree level (Associates vs Doctorate); different practice (RT is under the direction of a physician vs PT is a direct access provider).

Lines 170-171:

"Additionally, the increasing role of exercise in maintaining lung and general CF health is of great interest and represents another domain that can be taken on by the CF RT."

While a CF RT may have an “interest” in exercise, they do not have the education, training and licensure to assess and prescribe exercise. PT does! If the CF RT “takes on another domain” for which they are not educated or licensed, they would be outside their scope of practice!

All exercise is not the same – it must be prescribed and dosed for intended effects, with knowledge of how all the body systems contribute and are impacted by movement. This is true for maintaining lung health, optimizing postural alignment to promote ideal breathing mechanics and organ function, improving strength, managing pain, mitigating urinary incontinence, etc.

**Please provide any feedback on the section: Essential Consultant, lines 222-294**

Line 224-225

“Essential consultants (Figure 1) are healthcare providers whose expertise is needed because of complications that are highly prevalent in pwCF (Supplemental Table 3) and for which there are better outcomes when managed by those with specific knowledge and expertise in CF” .

The impairments that PT evaluates and treats falls under this category of provider based on your definition, as opposed to the Trained and Trusted Colleagues category. Trained CF

PTs have specific knowledge, as noted previously in comments for “Specialized CF Care Center”, and deserve to (at very least) be listed in the Essential Consultant category. However, I do believe there is enough evidence to support PT being a Core Team member. Please refer back to comments already provided in “any major topic the authors missed” regarding the CF PT having a network of PT specialists, much in the way the CF physician relies on their colleagues in other areas of medicine. The CF PT has knowledge about the whole person with CF, can intervene to their level of expertise, then refer to their PT colleagues in pelvic health, orthopedics, vestibular rehab, etc. as needed.

Line 266

“Dysphonia and hearing loss...”

Consider adding vestibular impairment.

**Please provide any feedback on the section: Trained and Trusted Colleagues, Lines 295-299**

Lines 296-297

“These providers have a fundamental understanding of CF, but the frequency at which they treat pwCF is not high.”

PTs are already established members of many care teams across the country, treating pwCF on a regular basis. PT does not belong in this category.

**Please provide any feedback on the section: Programmatic Components, lines 300-320**

**Please provide any feedback on the Conclusion, lines 321-351**

Line 324-325:

“...there is no evidence that the health needs over the course of the CF lifespan will decrease... suggesting that the existing CF team structure should be retained.”

Since PTs are already integral members at many CF centers, there is not rationale for the “demotion” to “Trained and Trusted Colleague”.

Line 328:

“Various models have been used to help integrate PCPs, GCs and pharmacists into healthcare team...”

Various models have also integrated PTs into their team. The CFF offers a grant to partially fund PTs in the CF clinic; this is a 3-year grant and is currently in its 3<sup>rd</sup> cycle. Since 2016, more than 70 programs have received funding for PT.

### **Do you have any feedback on the tables and figures?**

#### **Figure 1: Structure of a CF Care Center**

Physical Therapists have a range of education and expertise, from entry-level DPT to Clinical Specialists/subspecialists--thus the CF Care Team PT also has an "essential consultants" network for referral/consultation--not the same as the Clinic PT, who is the key person to coordinate the consultants and "trained and trusted colleagues" (e.g. personal trainers, athletic trainers, exercise physiologists)

Physical Therapists are members of Rehabilitation Services in most settings. This figure reflects a lack of knowledge about the profession, likely due to lack of representation on the writing group.

#### **Figure 2: Essential tasks conducted by the CF Core Team outside of clinic visits**

#### **Table 1: Trained and Trusted Colleagues**

The brief description of the roles for the Physical Therapist are not reflective of the conditions currently addressed by Care Center PTs and is quite incomplete regarding the effect of CF on all components of human movement across the lifespan. Many, if not all, of the emerging issues discussed in this document are impacted by physical activity and exercise, not just musculoskeletal issues and pain.

CF is much more than a lung disease, and PTs are qualified to address all issues as they impact movement/exercise

Removing need for routine ACT does NOT remove PTs value!!!

Exercise capacity is a better predictor of function than lung function or BMI--involves many more body systems than just respiratory and digestive--PT is expert in all the systems

HEMs improve lung function but not exercise capacity, showing that exercise requires more than just lungs! (analogy: taking anabolic steroids and sitting on the couch won't lead to being a champion weight-lifter)

Increasing BMI does not equate to better health--body composition is more important  
(which involves exercise)

**Supplemental Table 1: Suggested Staffing Ratios**

**Supplemental Table 2: Models for Integrating Primary Care Providers, Genetic Counselors,  
and Pharmacists into CF Care**

**Supplemental Table 3: Common Complications of CF that require Core Team or Essential  
Consultant care**

PT can help prevent and/or treat many of the complications listed: arthritis/arthropathy, osteopenia, osteoporosis, asthma, distal intestinal obstruction syndrome (bowel massage, defecation posture), anxiety disorder and depression.

Complications not listed here due to not being asked about on the registry data used for the 2022 Annual Report, but likely just as, if not more, common than what's listed: pain, urinary incontinence (recently added as an item on the registry), and vestibular dysfunction. All of these complications can be screened for and treated by a CF PT; the CF PT can refer to their specialist PT colleagues as needed for further evaluation and treatment.