

Improved Prognosis in Cystic Fibrosis: Consideration for Intensive Care During the COVID-19 Pandemic

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Letter to the Editor: Correspondence Letter

The dramatic improvement in prognosis for individuals with cystic fibrosis (CF) must not be overlooked in deliberations about the use of intensive care during the COVID-19 pandemic.

Thanks to advances in treatment options and improvements in care, the median predicted survival for people with CF in the US is approaching 50 years of age and steadily climbing.¹ With the recently approved combination modulator, elexacaftor/tezacaftor/ivacaftor (Trikafta™), which addresses the basic defect in CF, many people with CF are experiencing transformational improvements in quality of life and lung function that will undoubtedly further increase survival.

For patients with advanced CF lung disease, the outlook has also dramatically improved. Prior to FDA approval of Trikafta, the median survival for patients with CF and FEV1 <30% predicted was shown to be over 6.5 years.² Recent studies reveal improving critical care outcomes, with unanticipated survival and functional recovery from respiratory failure precipitated by influenza and other acute infections. These data prompted the 2020 CF Foundation consensus guidelines for the care of individuals with advanced CF to recommend that individuals be considered eligible for intensive care.³

As states and hospitals turn to emergency triage plans to ensure that scarce medical resources are allocated wisely, we are concerned that some plans use the mere presence of existing health conditions, including CF, as a determining factor in these decisions. This approach fails to recognize that CF has a wide range of disease manifestations. As importantly, categorizing CF in the same manner as other chronic lung diseases fails to consider that individuals with CF typically are decades younger and more functional relative to others with more common chronic lung diseases. While it is important to give frontline healthcare workers guidance during the crisis, considering all chronic lung diseases as similar, both as a risk factor for higher mortality from COVID-19 pneumonia and rationale for denying life-saving intensive care to people with CF, is too simplistic.

Alarming, several states have created regional triage plans that reflect an outdated understanding of CF and run the risk of denying care without taking current clinical realities into account. For example, Tennessee includes the presence of CF with FEV₁ of less than 30 percent as a factor for denying hospital admission.⁴ While risk-stratification is important when allocating resources, the blanket exclusion of individuals with CF and FEV₁ <30% predicted is based on an inaccurate understanding of the current survival outcomes for the CF patient population and does not factor in the short and long term impact of disease-modifying CF therapy.

The University of Pittsburgh has proposed an algorithm that looks beyond the presence of an underlying condition with the application of a multi-principle allocation framework to aid providers in these difficult times.⁵ This algorithm utilizes life expectancy of less than 5 years, prior to COVID-19 infection, as an indicator of lower priority for critical care or ventilator utilization. Individuals with CF and FEV₁ <30% predicted have a predicted median survival that is longer than 5 years and that data pre-dates the widespread availability of a transformational therapy, Trikafta. Accordingly, individuals with advanced CF lung disease should not have a lower priority for intensive care.

We are in unprecedented times and healthcare teams may face tremendously difficult decisions related to rationing ventilators and offering intensive care. As states and institutions consider revising existing triage plans or formulating new ones, decisionmakers should be careful to avoid language that excludes patients from receiving care because of an underlying condition without careful attention to the prognosis for those individuals. It is vitally important that crisis standards of care factor in accurate, disease-specific prognostic information as patients are triaged.

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