Cystic Fibrosis

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The advent of cystic fibrosis transmembrane conductance regulator (CFTR) modulator treatment has ushered in a new era of cystic fibrosis (CF) care. The impact of these therapies on survival, patient-reported outcomes, and quality of life is evolving and the configuration of CF care must adapt accordingly. This edition of Seminars in Respiratory and Critical Care Medicine delivers an up-to-date review of important areas of CF management in this new era of CFTR modulator treatment and provides guidance on expected modifications to clinical care, including a chapter dedicated to current status and evolving knowledge of these novel therapies.

Polymicrobial lung infection is a frequent complication of CF and several chapters focus on this area and the antimicrobial strategies used to combat these infections.

Viral exacerbations are a cause of significant morbidity and can hasten lung function decline in CF. A chapter is dedicated to this notable subject and is of particular relevance in a post-COVID (coronavirus disease) world. The use of digital technology in CF clinical care accelerated during the coronavirus disease 2019 (COVID-19) pandemic, and one article provides a comprehensive review of the current impact of digital technologies on clinical care for adults with CF.

Current CFTR modulators eligibility is dependent on CF genotype and, although it is predicted that lung transplant rates will continue to decline in the future, a significant proportion of people with CF (pwCF) who are not eligible for CFTR modulators will potentially need this rescue treatment. A manuscript addresses lung transplantation in CF in the age of CFTR modulators.

Median survival age is increasing in many countries and healthy aging in pwCF is a recognized future challenge for CF caregivers. Adaptation of physiotherapy regimens, disease monitoring, care delivery, and CF care beyond the lungs are tackled in this publication.

The future of CF care is bright; however, many challenges remain, and this edition of Seminars in Respiratory and Critical Care Medicine in CF is a well-timed account of the current and future landscape of CF management.