



Cystic fibrosis lung disease: from basic research to clinical issues

Guest Editor



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Message from the Guest Editor

Dear Colleagues,

Cystic fibrosis (CF) is the most common autosomal recessive diseases found within the Caucasian population, whose median survival has been increased due the introduction of novel therapeutic approaches. Although it is a syndrome, the main morbidity and mortality of CF people is caused by the lung disease. The lung involvement is usually progressive with intermittent exacerbations. CF is caused by mutations in the CF Transmembrane Conductance Regulator (CFTR) gene, a chloride channel, involved in the regulation of ion and fluid transport across different epithelium-lined organs. The genetics of CF is quite complex, with genotype-phenotype correlations depending on other genetic and environmental modifiers. The lack/defect of the CFTR protein leads to a dehydrated mucus, impaired mucus clearance, determining the formation of endobronchial mucus plaques and plugs, which become the main sites of air flow obstruction, infection, and inflammation. The immune



system, both innate and adaptive, seems to be primarily deranged, contributing to this pathophysiological process. The CFTR protein has been also involved in as many as different processes, such as foetal development, epithelial differentiation/polarization, and regeneration, as well as in epithelial-mesenchymal transition occurring during wound healing and cancer. In this issue, basic research and translational issues will be considered, including genetics of CFTR and other ion channel/transporters, aspects related to inflammation and immunity, as well as tumorigenesis. The identification of novel biomarkers of the lung disease that are specific, can identify CF children at risk for more progressive lung disease and serve as outcome measures for clinical trials is another intensive field of investigation. Finally, current therapeutic modalities, novel pharmacological and genetic therapies targeting the basic defect, cell-based therapies, and lung transplantation will be covered.

Prof. Massimo Conese and Prof. Lorenzo Guerra

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Submission Deadline: **31 December 2021**

Submission: <https://www.fbscience.com/Landmark>

Science Citation Index Expanded: **2.747** (2019)

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