

Cystic Fibrosis

*Infection, Immunopathology,
and Host Response*



Edited by

Richard B. Moss

Cystic Fibrosis Infection Immunopathology And Host Response

S. Jane Flint



Cystic Fibrosis Infection Immunopathology And Host Response:

Cystic Fibrosis Richard B. Moss, 2012-12-06 This work is concerned with Cystic Fibrosis CF the most common fatal genetic disease in the Caucasian population The decade of the 1980s was one of spectacular progress in understanding the genetic and molecular basis of CF The research breakthroughs of the decade began with the first fundamental insights published in 1981 1983 into the basic cellular pathophysiology of CF with demonstrations of altered ion transport in specialized exocrine epithelial tissues 1 3 Research progress shifted into a triumph of reverse genetics using restriction fragment length polymorphism DNA technology 4 with the localization of the CF gene to a region of chromosome 7 5 7 Understanding accelerated by an explosion of in vitro methodologies for epithelial cell culture and transformation allowed and physiological studies 8 11 these focused controlled biochemical with increasing precision on the molecular pathology of distal steps in the regulatory pathways for epithelial ion transport 12 19 Finally the end of the beginning occurred in late 1989 with one of the great achievements of molecular genetics the isolation and cloning of the CF gene 20 As a result we now have a CF gene product the cystic fibrosis transmembrane regulator CFTR possessing predicted amino acid sequence suggested tertiary structure and possible transmembrane transport function 21 These amazing developments have set the stage for the next round of advances which surely will include 1

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The Metabolic Basis of Inherited Disease John Bruton Stanbury, John B. Stanbury, 1983

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