

综述

CFTR与囊性纤维化

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摘要

囊性纤维化跨膜传导调节因子(CFTR)是一种cAMP激活的ATP门控性氯离子通道,表达于气道,消化道和生殖道上皮细胞的顶部质膜中。囊性纤维化(CF)是白人中最常见的遗传性疾病之一,由CFTR基因突变造成。对CFTR基因的破译使人们进一步了解CF的发病机制,并为该疾病的诊断提供了新的线索。

关键词 [囊性纤维化](#) [囊性纤维跨膜电导调节因子](#) [ATP结合盒](#) [ΔF508](#) [基因疗法](#) [药物治疗](#)

分类号

CFTR and cystic fibrosis

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Abstract

The cystic fibrosis transmembrane conductance regulator (CFTR) is a cAMP-activated and ATP-gated Cl⁻ channel expressed in the apical plasma membrane of epithelial cells in the airways, digestive and reproductive tracts. Cystic Fibrosis (CF), caused by mutations in the CFTR gene, is one of the most common inherited disorders of white populations. The identification of the CF gene led us to a further understanding of the CFTR structure and function, the mutational basis as well as the complexity of the disease.

Key words [cystic fibrosis\(CF\)](#) [cystic fibrosis transmembrane conductance regulator \(CFTR\)](#) [ATP binding cassette\(ABC\)](#) [deltaF508](#) [gene therapy](#) [drug treatment](#)

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