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Microbiological environment in the lower airway of the infants and young child with cystic fibrosis

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Cystic fibrosisis an autosomal recessive genetic disorder characterized by multi organ involvement and substantial heterogeneity in the disease phenotype. At a molecular level, the pathogenesis of CF is attributable, at least in part, to over 1800 known mutations within the cystic fibrosis transmembrane conductance regulator (CFTR) gene. CFTR is a necessary protein which allows normal transport of chloride and water across the cell surface. Genetic mutation analysis of the CFTR gene can delineate the severity of protein production, and may predict disease severity. Lung disease is the major cause of morbidity and mortality. The vast majority (85%) of CF patients carry two "severe" (pancreatic insufficient) alleles of CFTR; the remaining patients have some residual function ("mild", pancreatic sufficient, mutations). Current breakthroughs in therapy of the CF patient include the approval of ivacaftor, a small molecule drug which targets the CFTR protein and potentiates its function at the salt (chloride) channel on the cell surface. This medication has only been approved for use in approximately 4% of patients, with the G551D gating mutation. Additional corrector-potentiator combination medications are in clinical trials. Potential long term effects of these medications may alter the lower airway environment and it's susceptibility to infection.

With the advent of universal newborn screening (NBS) in the United States for cystic fibrosis, the age of diagnosis for the majority of infants is within the first month of life. Aggressive treatment of infants after NBS diagnosis confirms that with nutritional support and early treatment of cough illness and infection, there is perhaps a delay in the onset of chronic, progressive lung disease. However inflammation and air-trapping have been shown to be present in the lower airway of young infants and children, with early radiographic findings on computerized tomography (CT scan) as well.The onset of chronic infection by mucoid *Pseudomonas aeruginosa* leads to alginate production and the switch from plantonic growth of organisms to biofilm production. The *Pseudomonas* organisms in the biofilm communicate by quarom sensing; they are able to evade normal host defense preventing eradication of the organisms in the lower airway. Pulmonary exacerbations reduce health related quality of life and result in accelerated pulmonary function decline and decreased survival. The incidence of pulmonary exacerbations appear to be relatively constant over the life of a CF patient. The current life expectancy of a child born with CF in the United states in 2012 is 37.5 years.

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