Editorial

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Editorial Note on Cystic Fibrosis

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Editorial

Cystic fibrosis (CF), also known as mucoviscidosis, formerly cystic fibrosis of the pancreas, is a hereditary metabolic disorder, a hallmark of the production of strong, sticky mucus that closes the respiratory tract and intestinal tract. Cystic fibrosis was not recognized as a separate disease until 1938 and was classified as a pediatric disease because the mortality of traumatized infants and children was high. By the mid-1980's, however, more than half of all victims of cystic fibrosis survived to adulthood as a result of brutal treatment.

Cystic fibrosis is a genetic disease that mainly affects people of European descent. It is estimated that one in every 2,500 to 4,500 live births to these figures is concentrated in people of northwestern European descent. It is very rare in African ancestors (about one in 17,000 born alive) and is very rare in Asian ancestry. The disease had long been known as a recurrence — that is, only humans inherited a genetic predisposition from both parents. The disease does not show up in heterozygotes — that is, those individuals who have one common copy and one copy with the genetic makeup involved. However, when both parents are heterozygous, they may expect that, on the basis of luck, one in four of their offspring will develop the disease. In 1989 the gene that caused cystic fibrosis was isolated. The gene, called a cystic fibrosis transmembrane conductance regulator, or CFTR, is located in chromosome 7 and encodes a protein code with the same name, designated CFTR.

Cystic fibrosis affects the function of the body's exocrine glands - eg, mucus-secreting and sweat glands - in the respiratory and digestive systems. Within the lung and intestinal cells, the CFTR protein transports chloride to the cell membrane and regulates other channels. These activities are important for the maintenance and repair of mucous membranes. Many cases of cystic fibrosis are caused by mutations associated with the production of the CFTR protein free of the amino acid phenylalanine. As a result, chloride and sodium ions accumulate inside the cells, thus absorbing fluid from the cells and causing dehydration of the mucous membranes that normally cover these areas. Strong, sticky mucus builds up in the lungs, clogs the bronchi and makes breathing difficult. This causes chronic respiratory infections, usually with Staphylococcus aureus or Pseudomonas aeruginosa. Chronic cough, recurrent pneumonia, and progressive decreased lung function are major symptoms of lung disease, the most common cause of death in people with cystic fibrosis.

Among the most promising treatments being investigated for cystic fibrosis is heredity. Genetic therapy first emerged as a potential treatment in 1990, when researchers successfully restored the function of the CFTR chloride channel in epithelial cells of enlarged lungs and airways that carried CFTR mutations. Researchers have used synthetic DNA to produce viral vectors that contain copies of the CFTR gene. These vectors are then transferred to enlarged cells, which later incorporate normal genes into their DNA. This success led to the first clinical trial of genetic therapy for cystic fibrosis in 1993. The same technology was used to insert the CFTR gene into a repetitive adenovirus that was then inserted into patients' noses and lungs. This initial study appeared to be effective at first, as an increase in CFTR protein expression was observed immediately after treatment. However, patients experienced more serious side effects, including pneumonia and symptoms of infection. Since the 1990s, genetic treatment for cystic fibrosis has greatly improved, and the results of clinical trials are marked by continuous improvement. However, the natural immune systems of the lungs and airways have proven to be major barriers to cellular viral retrieval of cells that carry the genetic CFTR gene. As a result, the development of an effective genetic delivery system has become increasingly focused on genetic treatment for cystic fibrosis. The delivery systems under investigation include cationic polymer vectors, cationic liposomes, and adenovirus-related virus. The latter, which could bind to the type of receptor expressed in high numbers in the upper extremities of the lung cells, proved to be particularly effective in laboratory research using human lung tissue.

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None

Conflict of Interest

None