**Introduction.** Cystic fibrosis is the most common autosomal recessive lethal hereditary disease with the multiple organ manifestation [5; 7; 23]. Prognosis and severity of cystic fibrosis to a great degree depends on the chronic infectious inflammatory process of bronchopulmonary system [4]. The crucial role in the progress of bronchopulmonary disease and death during cystic fibrosis belongs to the chronic Pseudomonas aeruginosa [3; 8]. In case of steady bacterial colonization of the respiratory tract by *Pseudomonas aeruginosa* the protective function is performed by non-specific factors of humoral immunity, such as antimicrobial peptides [6; 41; 51]. An optimizing influence on functioning of non-specific mechanisms of the immune response due to increase of production and expression of antimicrobial peptides is shown by vitamin D [27; 52]. Resistance of *Pseudomonas aeruginosa* to antibiotics is one of the main problems in treatment of patients with chronic infections of the respiratory organs during cystic fibrosis. For this reason, a particular interest is now attracted to the research and use of alternative versions of common antibiotics in treatment of cystic fibrosis – antimicrobial peptides [29; 54], as well as substances inducing their production and expression [25].

*Cystic fibrosis, CF* – a hereditary systemic exocrine disorder, caused by mutation of gene *CFTR* (cystic fibrosis transmembrane conductance regulator) and characterized by autosomal recessive type of inheritance.

**Epidemiology of cystic fibrosis.** Frequency of cystic fibrosis in different geographical regions of the world varies from 1:600 to 1:350 000 births [53]. In the European countries this disease occurs, on the average, once in every 2 000 – 3 000 births [23; 53]. In Ukraine frequency of cystic fibrosis is one patient in every 3 364 births [7].

**Mutation of gene CFTR.** According to the CFTR Consortium data 2014, there are 1 995 various CFTR gene mutations, in 177 of cases clinical manifestations of cystic fibrosis develop [32]. As a result of multicentre research in the European countries, 26 most common mutations have been defined [50].
Clinical peculiarities of cystic fibrosis, associated with the CFTR gene mutation type. Association of the specific CFTR mutation with the disease severity depends on the mutation type, its position inside the gene and influence on the structure and function of transmembrane regulatory protein cystic fibrosis [24; 32].

Peculiarities of bacterial contamination of mucous membrane of bronchial tree in chronic bronchitis of children with cystic fibrosis. Pathologic changes in cystic fibrosis of all the excretory glands are caused by the failure of transportation process of chlorides through the membranes of epithelial cells. Adhesive bronchial mucus blocks the movement of pathologically changed cilia of bronchial epithelium, and mucus components easily flocculate. As a result of these changes, bronchi self-cleaning mechanism becomes violated, which induces joining of pathogenic flora and development of inflammation [38]. According to the multicentre international research, Pseudomonas aeruginosa is a pathogen playing the crucial role in the progress of bronchopulmonary disease and death in cystic fibrosis [45].

The role of CFTR in internalization of Pseudomonas aeruginosa by epithelial cells of the airway. Besides mucociliary clearance, it is considered that Pseudomonas aeruginosa are removed from the bronchopulmonary system after the internalization of epithelial cells and the ensuing deletion of cells of the airway containing pathogen. CFTR is a receptor for internalization of Pseudomonas aeruginosa, so insufficiency of CFTR function may change the bacterial contamination in bronchi and lungs in patients with cystic fibrosis [38; 41].

Significance of antimicrobial peptides in the chronic infectious inflammatory process of respiratory organs in cystic fibrosis. In the respiratory tract neutrophils in the presence of microorganisms express agents which have antibacterial activity, such as cathelicidin (LL-37) and α-defensins. In their turn, epithelial cells of the respiratory organs produce β-defensins []. These antimicrobial peptides demonstrate expressed bactericidal activity against Staphylococcus aureus, Haemophilus influenzae and Pseudomonas aeruginosa [1].
Sputum of children with cystic fibrosis has high content of neutrophils and antimicrobial peptides with the steady bacterial colonization of the respiratory tract [6; 41].

*Mechanisms of vitamin D-dependant production of antimicrobial peptides in the respiratory tract.* In the research by Tian-Tian Wang et al. [52] it was stated that during activation of Toll similar receptors (TLR) of epithelial cells of the respiratory tract calcitriol (1,25-Dihydroxy cholecalciferol - 1,25(OH)2D) induces expression of β-defensins and cathelicidin in monocytes and neutrophils. Moreover, 1,25(OH)2D induces the corresponding production of antimicrobial peptides and activation of antimicrobial activity in reference to gram-negative pathogens, such as *Pseudomonas aeruginosa*.

*Clinical significance of vitamin D-dependant production of antimicrobial peptides in cystic fibrosis.* During the last decade a number of researches has increased aimed at study of clinical laboratory efficiency of the use of vitamin D in treatment of patients with cystic fibrosis with infectious bronchopulmonary complications [13; 18; 25]. The researches show that replacement therapy with high doses of cholecalciferol (vitamin D3) (100 000 ME-600 000 ME/2500-15 000 mkg) on a once-only basis inward or intramuscularly is a more effective method of treatment vitamin D deficit conditions in cystic fibrosis and has received a name “Stoss therapy” [21].

*Conclusion.* The research makes it possible to conclude that chronic infection of the airway of children with cystic fibrosis affects the course of disease due to the natural resistance of *Pseudomonas aeruginosa* to the antibacterial medication. Antimicrobial peptides have a high antimicrobial activity against gram-negative bacteria in the respiratory tract. Capacity of vitamin D to increase production of antimicrobial peptides allows to recommend it as a medicine in complex therapy of chronic infection of respiratory organs in cystic fibrosis.