EUROPEAN RESPIRATORY *journal*
FLAGSHIP SCIENTIFIC JOURNAL OF ERS

Questions of predicting and preventing the progression of pneumofibrosis in children with cystic fibrosis

Svetlana Ilchenko, Anastasia Fialkovska, Svetlana Ivanus
European Respiratory Journal 2019 54: PA966; **DOI:** 10.1183/13993003.congress-2019.PA966

Article | Info & Metrics

Abstract

Inflammation of the respiratory tract in patients with cystic fibrosis (CF) occurs at the beginning of the disease, manifests itself even in the absence of infection. Therefore, the use of antibiotics is not sufficient to stop the inflammation. The accumulation of sputum with high content of DNA contributes to the development of a large amount of transforming growth factor (TGF-β). TGF-β is the mediator of fibrosis, the rate of disease progression and pulmonary complications.

The aim is to study the role of Dornase alpha in the prevention of pulmonary fibrosis in children with CF.

Methods: 47 children with CF were examined, among them patients without respiratory symptoms and with chronic bronchitis. TGF-β was determined in the endothelial tissue of the bronchi and in the blood serum.

Results: TGF-β in endothelial bronchial tissue was identified in the quarter of children. The intensity of expression of TGF-β was more pronounced in children without chronic bronchitis. This is a prognostic criterion for the development of "inadequate" pneumofibrosis. In children with positive TGF-β expression in the bronchial endothelium the level of this marker in the blood was significantly higher. It

was found that the treatment with Dornase alpha for 4 weeks significantly lowered TGF-β levels in children by 12 % during the exacerbation of the disease.

Conclusion: The inclusion of Dornase alpha in the treatment of patients with CF should begin even in the absence of respiratory symptoms. Determination of serum TGF-β levels in children with CF can help predict the severity of the course and select an optimal therapeutic strategy that will slow down the progression of the disease.

Children Cystic fibrosis Inflammation

Footnotes

Cite this article as: European Respiratory Journal 2019; 54: Suppl. 63, PA966.

Previous

Back to top

Vol 54 Issue suppl 63 Table of Contents

Table of Contents
Index by author

Email
Alerts
Citation Tools

© Request Permissions
Share

Jump To

Article
Info & Metrics