

Cystic Fibrosis Research News

Title:

YKL-40 as marker of severe lung disease in Cystic Fibrosis patients

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What was your research question?

The aim of this study was to evaluate serum (the clear yellowish fluid obtained upon separating whole blood into its solid and liquid components after it has been allowed to clot) and sputum levels of a protein called YKL-40 in cystic fibrosis patients, to examine the relationship between serum and sputum YKL-40 levels with disease severity.

Why is this important?

YKL-40 is a protein belonging to “Chitinases family”, enzymes characterized by their ability to split chitin. Chitin is a carbohydrate (‘sugar’) or structural protein used by a variety of organisms including insects, crustaceans, parasites, fungi and bacteria to make their exoskeletons and protect against harsh environmental conditions. In healthy humans YKL-40 is produced by neutrophils, monocytes, macrophages, which are types of white blood cells that regulate cell growth and are essential, as elements of the immune system, to defend the host from infection by other organisms. YKL-40 levels have been shown to increase in some severe human diseases, such as cancer, osteoarthritis, cardiovascular diseases, neurological diseases and injury, infections, chronic obstructive pulmonary disease, asthma and recently even in cystic fibrosis adults.

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What did you do?

Serum and sputum YKL-40 levels were measured in a group of twenty-eight patients with a diagnosis of cystic fibrosis and twenty healthy people to act as controls.

What did you find?

We demonstrated that cystic fibrosis patients are characterized by higher levels of both serum and sputum YKL-40 when compared with healthy subjects. Furthermore in cystic fibrosis patients older than 18 years we found both in serum and in sputum significantly higher YKL-40 levels than in cystic fibrosis patients younger than 18 years, confirming the relationship between this marker and the progression of the disease. When comparing YKL-40 levels between CF patients with different mutations, we observed that those carrying the most prevalent genetic mutation called $\Delta F508$, had YKL-40 levels that were not significantly higher than in patients with other different mutations, confirming that YKL-40 levels in cystic fibrosis patients are not the direct result of the genetic cystic fibrosis defect.

What does this mean and reasons for caution?

This study provides evidence that the levels of this protein are linked to disease severity. This could be useful for clinicians and to have a parameter to evaluate the severity and the progression of the disease.

What's next?

Longitudinal studies (observational research method in which data are gathered for the same subjects repeatedly over a period of time) in infant are needed to establish if YKL-40 levels are not only important as a proxy to gauge disease severity but also if are themselves the cause of the disease severity.

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