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Cystic Fibrosis Research News

Title:

Real-world assessment of LCI following lumacaftor-ivacaftor initiation in adolescents and adults with cystic fibrosis

Lay Title:

Use of LCI to monitor CFTR Modulator effect in people with CF

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What was your research question?

Can LCI be useful to monitor Lumacaftor-ivacaftor effect in people with CF (pwCF) in real world ?

Why is this important?

LCI has become very popular for monitoring lung function of pwCF. Until now, it has mainly been used in clinical trials with CFTR modulators tested in young children with CF with mild lung disease. Data are scarce on its usefulness in real world for monitoring CFTR modulators effects in older children and adults with CF.

What did you do?

In the present study, we took advantage of LCI measurements performed in a large national real-world observational study that followed CF patients aged 12 years or older over the first year after lumacaftor-ivacaftor (Lum-Iva) initiation. Sixty three out of 845 patients had LCI measured before, 6 and 12 months after Lum-Iva initiation.

What did you find?

At inclusion, pwCF were mainly adolescent. They had moderate lung dysfunction as measured by usual lung function test (in our study FEV1 measurement). Their LCI before starting Lum-Iva was elevated. After 6 and 12 months of treatment with Lum-Iva, there were no changes in both usual lung function test and LCI. Discordant results between LCI and FEV1 were observed in one-third of our patients, meaning, for instance that for some individuals, FEV1 was “improving” while LCI was “deteriorating” under treatment.

What does this mean and reasons for caution?

At the group level, in people with more advanced lung disease, we did not observe statistically significant improvements of lung function parameters under Lum-Iva. In this population, results were more heterogenous than those reported in children with milder disease.
For an individual treated with Lum-Iva, discordance was seen in around one patient out of three which makes the interpretation of results tricky for clinicians. Both tests brings different informations on lung health. They do not measured the same components of the CF related lung disease. This may be part of the reason for discordance.

What’s next?

LCI remains a promising lung function test for monitoring CFTR modulators in people with mild lung disease. Other observational studies will be needed to gain insights real world data

evolution under more efficient CFTR modulators prescribed in younger patients with milder lung disease.

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