European Cystic Fibrosis Society

Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

Cystic Fibrosis Research News

Title:

Real-life acute lung function changes after lumacaftor/ivacaftor first administration in pediatric patients with cystic fibrosis

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What was your research question?

Is the lumacaftor/ivacaftor combination (LUM/IVA) responsible for bronchoconstriction (airway narrowing due to airway smooth muscle contraction) in adolescents with cystic fibrosis after its first administration?

Why is this important?

Administration of LUM/IVA in healthy people has been consistently reported to induce bronchoconstriction associated with a mean drop of 4.1 points in the lung function parameter FEV_1 (forced expiratory volume in one second). Respiratory manifestations such as cough (n = 2) or shortness of breath (n = 1) have been reported in 26 patients. Bronchodilator inhalation totally restored FEV_1 loss.

What did you do?

We evaluated FEV_1 four hours after the first administration of LUM/IVA in a population of 32 adolescents with a mean age of 15.5 years and a normal baseline FEV_1 of 90%. We also recorded the occurrence of any respiratory manifestations. Finally, we studied the change in FEV_1 after administration of a short-acting bronchodilator through a spacer.

What did you find?

The first administration of LUM/IVA consistently induced a decrease in FEV_1 by a mean of 10 percent, but only three of the 32 adolescents had associated mild clinical manifestations (wheezing spells). Bronchodilator inhalation only allowed for a partial recovery of FEV_1 loss.

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We found that patients with low lung function at baseline and a past history of significant reversible airway obstruction (defined by an increase in FEV_1 of more than 12% after bronchodilator inhalation) were more at risk of having a steeper FEV_1 drop.

What does this mean and reasons for caution?

Patients with a known history of significant reversible airway obstruction (i.e., ≥12%) and a low baseline FEV_1 are more at risk of experiencing a steep FEV1 drop and should be closely monitored in clinics for a few hours after the first LUM/IVA administration.

What's next?

Studying how a drop in FEV_1 due to bronchoconstriction can be prevented by long-acting bronchodilator administration before the first LUM/IVA administration.

Original manuscript citation in PubMed

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