Title:
Continuous glucose monitoring in youth with cystic fibrosis treated with lumacaftor-ivacaftor

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What was your research question?
Do blood sugars in patients with cystic fibrosis, measured by continuous glucose monitoring, improve after starting lumacaftor/ivacaftor?

Why is this important?
Previously, a small study found improvements in insulin secretion and diabetes for patients with CF and G551D mutations after starting ivacaftor treatment. The effects of lumacaftor/ivacaftor (Orkambi) on blood sugars in patients with F508del mutations are unknown. As newer CFTR modulators are developed for different mutations, it is important to perform ongoing assessments of the effects of these medications on blood sugars and CF-related diabetes.

What did you do?
Nine individuals, ages 11-15 years, with homozygous F508del mutations wore a continuous glucose monitor (CGM) before and after Orkambi treatment. One had CF related diabetes, 5 had abnormal glucose tolerance tests, and 3 had normal glucose tolerance tests prior to Orkambi. CGM data were collected and compared roughly a median of 26 weeks before and 29 weeks after Orkambi initiation.
Cystic Fibrosis Research News

What did you find?
There were no significant differences in blood sugars measured by continuous glucose monitoring before vs after Orkambi treatment in this small group of people. In fact, one individual with normal glucose tolerance before Orkambi developed diabetes after Orkambi start.

What does this mean and reasons for caution?
Blood sugar abnormalities in the F508del population may be harder to correct with Orkambi than in the G551D population treated with ivacaftor. However, in this study, insulin secretion problems, which are the underlying cause of blood sugar abnormalities in diabetes, were not directly measured.

What’s next?
Future studies measuring both insulin secretion and blood sugar changes, before and after the start of newer CFTR modulator drugs, are needed to better understand the impacts of these medications on blood sugars, risk for diabetes, and diabetes progression.

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