

Cystic Fibrosis Research News

Title:

Abnormal Preschool Lung Clearance Index (LCI) reflects clinical status and predicts lower spirometry later in childhood in Cystic Fibrosis.

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What was your research question?

In this study, we investigated the utility of Multiple Breath washout (MBW) in the preschool age range (defined internationally as age 2-6 years). MBW assesses how even gas mixing within the lungs is. The most common result reported from the test is called the Lung Clearance Index (LCI): the higher the LCI the more abnormal the gas mixing is. We looked at Lung Clearance Index (LCI) results during the pre-school age range, how it related to measures of health status at the time of testing, and its relationships to later lung function results (spirometry) to better understand the clinical usefulness of LCI.

Why is this important?

Early detection of cystic fibrosis (CF)-related lung disease remains an important aspect of efforts to further improve CF-related outcomes. Multiple breath washout (MBW) is an easy to perform technique where the child breathes oxygen during normal relaxed breathing whilst being distracted by a movie/show. It can detect early obstructive lung disease in these young children. It is not part of routine CF clinical care yet. To help this happen, we need to better understand how it reflects clinical status and its ability to predict more established future health outcomes based on measures such as spirometry, the current gold standard for lung function monitoring. There is limited experience of MBW's utility in this setting using current commercially available equipment.

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What did you do?

We have been using MBW clinically for several years and looked back at the LCI results of the initial MBW tests on 50 of our preschool children within the CF clinic at the Children's Hospital at Westmead from 2014 to 2018. MBW had been performed using commercially available equipment. We looked at clinical data from the time of testing, such as CF genotype (mutation type), admissions to hospital, how much medication they had been prescribed (e.g. antibiotics, dornase alfa) and whether we had seen a lot of bacteria in their sputum samples up to that point. We also looked at their spirometry results later on, on average over 2 years later.

What did you find?

LCI was abnormal in the majority of preschool children tested (56% of them). Higher (more abnormal) LCI was associated with higher medication use (e.g. dornase alfa use) and a more severe CF genotype (pF508.del homozygous genotype – the most common mutation). Some analyses also suggested an association with greater numbers of previous positive cultures to bacteria, such as staph aureus. Abnormal initial preschool LCI was associated with lower later spirometry and importantly LCI was the strongest preschool predictor of low future spirometry outcomes.

What does this mean and reasons for caution?

The results show that MBW performed with commercial based equipment, which is widely available to interested CF clinics, contains information that is of clinical value. Results obtained were related to both current and future health status in CF subjects. LCI during the preschool age range was related to other current measures of concerning health status and contained information about how likely future lung function would be when performed with spirometry. This growing evidence with recent commercial devices strengthens the argument for routine preschool screening in children with CF, providing adequate training can be provided to ensure high feasibility and strong data quality.

What's next?

Future studies need to outline how best to use the MBW in these young children to improve CF outcomes. For example, which LCI values to act on and what treatments improve LCI in this age range if abnormal. This information is vital to help integrate MBW into routine clinical care.



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