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Cystic Fibrosis Research News

Title:
Lung structural and functional impairments in young children with cystic fibrosis diagnosed following newborn screening – a nationwide observational study

Lay Title:
Lung function and imaging outcomes in children with cystic fibrosis diagnosed after newborn screening

Authors:
Bettina S. Frauchiger1*, Corin Willers1,6*, Jasna Cotting 1, Elisabeth Kieninger1, Insa Korten1, Carmen Casaulta1, Yasmin Salem1, Enno Stranzinger2, Ben Brabandt2, Jakob Usemann3,4, Nicolas Regamey5, Alena Kuhn6, Sylvain Blanchon7, Isabelle Rochat7, Grzegorz Bauman8, 9, Dominik Müller-Suter6, Alexander Moeller3, Philipp Latzin1#, Kathryn A. Ramsey1,10# and the BILD and SCILD study groups

* Authors contributed equally to first authorship
# Authors contributed equally to senior authorship

Affiliations
1. Pediatric Respiratory Medicine, Department of Pediatrics, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland
2. Diagnostic, interventional and pediatric radiology, Department of Pediatrics, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland
3. Division of Respiratory Medicine and Children's Research Center, University Children's Hospital Zurich, Zurich, Switzerland
4. University Children's Hospital Basel (UKBB), Basel, Switzerland
5. Department of Respiratory Medicine, Children's Hospital Luzern, Luzern, Switzerland
6. Department of Paediatrics, Kantonsspital Aarau, Aarau, Switzerland
7. Hôpital de l'enfance de Lausanne, Lausanne Switzerland
8. Division of Radiological Physics, Department of Radiology, , University of Basel Hospital, Basel, Switzerland
9. Department of Biomedical Engineering, University of Basel, Allschwil, Switzerland
10. Wal-yan Respiratory Research Centre, Telethon Kids Institute, University of Western Australia, Perth WA Australia

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What was your research question?

We studied novel lung function and magnetic resonance imaging (MRI) outcomes in preschool and early school age children with cystic fibrosis that were diagnosed after newborn screening in Switzerland

Why is this important?

Newborn screening allows for detection of cystic fibrosis shortly after birth before children become clinically symptomatic. Therefore we aimed to assess whether this influences their lung function and lung disease later in life, at early school age. We also hypothesized that lung diseases may be milder from what we experienced in clinical routine and assessed whether conventional examinations are sensitive enough or novel parameters perform better.

What did you do?

We invited all children born after 2011 that were diagnosed by newborn screening with cystic fibrosis across Switzerland to a study visit at preschool/early school age in our centre (Bern). At this visit we measured lung function including conventional spirometry and the novel technique multiple breath washout. Multiple breath washout measures how long it takes air to distribute within the lungs and is known to be very sensitive for diseases affecting the very periphery of the lungs. Further, we performed lung magnetic imaging, which does not need sedation nor contrast agents and is without any radiation. Here we aimed to study how the lung structure but also the function in terms of distribution of air is affected. We also assessed the influence of clinical risk factors such as bacterial colonization, respiratory symptoms and pulmonary exacerbations.

What did you find?

We found that in general, lung disease in children with CF diagnosed after newborn screening is mild. Then we could also show that traditionally used spirometry is not different between healthy controls and CF children. In contrast, outcomes from multiple breath washout was already abnormal in ~20% of the children and MRI showed structural and functional abnormalities in 47%. This indicates that these measures are better to capture the mild lung disease in young and early-diagnosed CF children that mostly do not have clinical symptoms. We could also show that children that do have respiratory symptoms early in life and experience pulmonary exacerbations have impaired lung function in later childhood.

What does this mean and reasons for caution?

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Our findings show that the introduction of newborn screening lead to generally mild lung disease in childhood. However, the portion of children with abnormal multiple breath washout and MRI outcomes highlights that it is important to capture those children to prevent further progression of the lung disease, as most of them do not even have clinical symptoms. Clinicians should also identify children with early respiratory symptoms as they have a greater risk of impaired lung function later in childhood. In addition, pulmonary exacerbations should be prevented or treated strictly as a known risk factor for lung function impairment.

What's next?

In a next step we aim to asses the influence of newly available modulator therapies in this age group and whether this leads to even better lung function and less structural lung disease.

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