INSELSPITAL
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SCILD Swiss CF Infant Lung Development
BILD Cohort Basel Bern Infant Lung Development Cohort

Early lung function tests in infants with cystic fibrosis

Background and aim of the study: In individuals with cystic fibrosis (CF) lung function deteriorates faster than healthy individuals. When exactly this deterioration takes place and whether lung function is abnormal in infancy is unknown.

Measurements: Lung function tests were performed during natural sleep in 53 children with CF (out of the SCILD cohort) and 57 healthy children (out of the BILD cohort) between 5 and 10 weeks of age.

Results: More than 40% of the infants with CF already had an impaired lung function. The lung clearance index (LCI), a measure of uneven gas mixing in the lung, and/or the functional residual capacity (FRC), a marker of hyperinflation, were elevated in 22 out of 53 children. The elevation of these two parameters was independent of clinical symptoms or earlier therapies. The results show that in some children with CF lung function is already impaired shortly after birth and before respiratory symptoms commence. Hence, lung function measurements could possibly be used in infancy to monitor early CF disease and/or response to therapy.

Publication: Kieninger et al.; Elevated lung clearance index in infants with cystic fibrosis shortly after birth. European Respiratory Journal 2017

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