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Title and Citation:

Improvements in lung function and height among cohorts of 6-year-olds with cystic fibrosis from 1994 to 2012. J Pediatr. 2014 Dec;165(6):1091-1097.

Authors:

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What was your research question? (50 words maximum)

We asked how lung function and height have changed among 6-year-old children (when we first record lung function) with CF followed in the CF Foundation Patient Registry (CFFPR) between 1992.

Why is this important? (100 words maximum)

Preserving lung function is an important part of maintaining CF health, and lung function begins to be lost at birth in CF. Average lung function has improved among children with CF followed in the CFFPR over the past two decades, but we don't know if that is because all types of children (i.e., both sexes, all races, all genotypes, all insurance types) are healthier today than children of the past, or if children with some backgrounds have benefited more than others. If we can identify groups that haven't benefitted as much, we can change our practices to help them more.

What did you do? (100 words maximum)

For each year from 1992 to 2012, we collected the best recorded lung function measures for children that were 6 years old during that year, along with background information (their race, sex, CF mutation type, insurance background, height, and how they had been diagnosed). Then we compared how average lung function had changed from year to year among all these children to how it had changed among children from different backgrounds.

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We compared changes in girls versus boys, changes in children with two F508del mutations to other children, and changes in children insured by Medicaid to those who were not.

What did you find? (100 words maximum)

Averages of two lung function measures, FEV1 and FVC, and height steadily and significantly improved in all children between 1992 and 2012. Improvements were seen among children regardless of background (sex, CF mutations, or insurance coverage). Children who had been diagnosed by CF newborn screening had significantly better lung function at age 6 than their peers who had not been, suggesting an important benefit of early nutritional intervention for children with CF. Although all groups improved between 1992 and 2012, some were healthier than others in 2012, suggesting room for greater intervention in very young children to improve lung health.

What does this mean and reasons for caution? (100 words maximum)

Our results suggest that changes in the care of young children with CF have produced an overall improvement in the health of 6-year-olds with CF, with the introduction of CF newborn screening associated with a large improvement. However, our results also suggest that there appears to be room for additional improvements in lung and nutritional health in children with CF. In addition, differences remain in the overall health of children with CF from different backgrounds, suggesting that some groups of children might benefit from greater attention and more rigorous interventions than they may currently receive.

What's next? (100 words maximum)

During the last year of this study (2012), only 31% of 6-year-olds had been diagnosed by CF newborn screening. Currently, greater than 90% of children

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are being diagnosed by screening. Repeating these analyses in the future may identify groups of children that would benefit from earlier intervention for whom more rigorous interventions may be needed.