

Cystic Fibrosis Research News

Title:

Health-related quality-of-life in children with cystic fibrosis aged 5-years and associations with health outcomes

Lay Title:

Quality of life and health outcomes in children with CF aged 5 years

Authors:

Joyce Cheney^{a, b}, Suzanna Vidmar^{c, d}, Nicholas Gailer^a, Claire Wainwright^{a, b}, Tonia A Douglas^{a, b}, for the Australasian Cystic Fibrosis Bronchoalveolar Lavage (ACFBAL) study group

Affiliations:

^aDepartment of Respiratory and Sleep Medicine, Queensland Children's Hospital, Brisbane, Australia

^bCentre for Children's Health Research, School of Medicine, The University of Queensland, Australia

^c Clinical Epidemiology and Biostatistics Unit, Murdoch Children's Research Institute, Melbourne, Australia

^d Department of Paediatrics, University of Melbourne, Melbourne, Australia

What was your research question?

We wanted to know if the health related quality of life (HRQoL) of children with CF at age 5 was: a) the same as healthy children who do not have CF b) related to children's CF health status and c) whether children rated their own HRQoL differently to how their parents rated their HRQoL.

Why is this important?

We do not know much about HRQoL in young children with CF and need to better understand the impact of CF health on wellbeing in young children. We want to know if measuring HRQoL might be a useful marker of CF physical health in children at this age. We don't know the differences between how children rate their own HRQoL and how parents rate their children: are parent reports reliable substitutes in younger children?

What did you do?

Children aged 5 years and their parents taking part in the Australian CF BAL study completed HRQoL questionnaires. The scores from the HRQoL questionnaires were compared to

Cystic Fibrosis Research News

published scores in healthy children. Scores among children with CF in this study were then related to their CF lung and nutritional health outcomes (lung function, weight, CT scans etc.). Children's self-reported scores were compared to their parent-reported scores to see how well they matched.

What did you find?

Children with CF at 5 years had worse HRQoL scores than healthy children who do not have CF. Parents tended to score their child's HRQoL lower (worse) than children did, and reports from parents are not substitutes for child-reports of HRQoL. Worse HRQoL was associated with worse lung function and nutrition, more lung exacerbations, and worse score on CT scans. This was the case only when using parent-rated HRQoL scores, but not the case when using child-rated QoL scores.

What does this mean and reasons for caution?

How parents rate their child's HRQoL is different to how children rate their HRQoL and it is important to measure both to understand children's experiences. HRQoL is lower than their healthy peers and CF teams need to focus on improving wellbeing in young children. HRQoL scores are associated with CF health outcomes, showing the close relationship between wellbeing and physical health in early life. HRQoL scores could be used in clinical trials as markers of success of new CF therapies. The HRQoL measure used was designed for children aged 6 years+ and this limits how reliable our findings are in 5-year-olds.

What's next?

We need a version of the HRQoL questionnaire designed for preschool children so we can understand their experiences and wellbeing. The factors that affect HRQoL in young children with CF need further investigation.

Original manuscript citation in PubMed

<https://pubmed.ncbi.nlm.nih.gov/32165156/>