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Cystic Fibrosis Research News

Title:

IMPACT OF A PROGRAM ENSURING CONSISTENT RESPONSE TO ACUTE DROPS IN LUNG FUNCTION IN CHILDREN WITH CYSTIC FIBROSIS

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What was your research question?

Can we create a systematic approach to ensuring that pulmonary exacerbations are diagnosed, treated, and followed up in order to better preserve lung function in people with cystic fibrosis (CF)?

Why is this important?

While antibiotic treatment of pulmonary exacerbations is a cornerstone of CF care, patient registry analyses show that many individuals with substantial acute drops in lung function go untreated, making it less likely for them to recover. Furthermore, CF care centers whose patients have above average lung function appear to treat pulmonary exacerbations (and, specifically, acute drops in lung function) more consistently and more frequently than the average center. It stands to reason that methods to ensure that pulmonary exacerbations are consistently treated will lead to better preservation of lung function in people with CF.

What did you do?

In 2012, we established a pulmonary care algorithm (process) which by default suggested the prescription of antibiotics in response to either drops in lung function or presentation of new signs and symptoms of pulmonary exacerbation, with consistent follow-up appointments to ensure recovery to baseline. We created tools to ensure that any drops in lung function would be highlighted during clinic visits. Patient trend reports and the goals of the visit were discussed during preclinic meetings, and shared with families, who were carefully integrated into the process. Data on each medical provider's adherence to the pulmonary care algorithm

and the average lung function of the clinic population was also tracked over time and displayed. Data for the period 2012 to 2017 were analysed.

What did you find?

Adherence by medical providers to various components of the algorithm improved over time from 32% to 85%, but was never 100%. This is fine as the intention of the algorithm was to provide guidance but not dictate all care decisions. Average lung function of individuals at our care center increased substantially as a result, from 87% to 98% predicted, and the gap in average lung function between adolescents 13-18 years of age and younger children 6-12 narrowed considerably as decline in adolescents was particularly impacted upon. Overall, the average lung function of our patients, which was in the lowest decile among US care centers in 2012, rose to the highest decile in the years 2015-2017.

What does this mean and reasons for caution?

Improvements in pulmonary outcomes can be accomplished rapidly using standardized and proactive approaches that ensure consistent recognition and antibiotic treatment of pulmonary exacerbations along with regular follow-up, and the use of data to follow the effectiveness of the process. While the Children's Hospital of Richmond is a single medium size pediatric CF program and results may vary at other programs, we believe that the steps involved would be easy for other CF Centers to adapt to their own settings. Adoption of this approach may lead to substantial gains in preservation of lung function in people with CF.

What's next?

Discussions are underway to introduce our approach at a large group of CF Centers participating in a quality improvement collaboration. We have also devised an algorithm to ensure a consistent response to low body mass index in those who attend our centre, and this has yielded substantial improvements in nutritional status of our population.

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