

Cystic Fibrosis Research News

Title:

Differences in clinical outcomes of paediatric cystic fibrosis patients with and without meconium ileus

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What was your research question?

Meconium ileus occurs in approximately 20% of newborns with cystic fibrosis due to abnormally thick and impacted meconium which blocks the small bowel.

Our research question was "Do paediatric cystic fibrosis (CF) patients with meconium ileus (MI) fare as well as those without MI?"

Why is this important?

There are varying and conflicting reports in the literature about the health outcomes of people with CF with MI. Some reports show these patients have worse health outcomes, but other reports found no differences. We wanted to evaluate the health outcomes of this population of patients as this may impact their management.

What did you do?

We looked back at the data of 930 CF patients in New South Wales (NSW), Australia, from 1988-2010 using the Australian CF Data Registry. We identified 162 patients who had MI and compared the clinical outcomes of patients with and without MI over time. These clinical outcomes were lung function, bacteria in airway samples, growth, nutritional interventions, gastrointestinal complications, CF-related diabetes, hospital admissions, transplants and deaths.



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What did you find?

CF patients with MI are more likely to have poorer lung function, poorer growth (body mass index) and increased nutritional support requirements (oral feed supplements and gastrostomy formation) compared to CF patients without MI. Having had MI was also associated with higher rates of hospitalisations (for gastrointestinal and respiratory reasons), transplants and deaths.

What does this mean and reasons for caution?

CF patients with MI were associated with worse health outcomes compared to those without MI. Closer monitoring and earlier aggressive management may be of benefit in this population. The study design was limited by its retrospective nature and the data restricted to a single state in Australia.

What's next?

Future long-term studies are required to confirm these findings.

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