



Cystic Fibrosis Research News

Title:

Baseline CF disease severity has an adverse impact on pregnancy and infant outcomes, but does not impact disease progression

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What was your research question?

Is there a relationship between CF disease severity and the course of the pregnancy? How does disease severity of women with CF affect the outcome of the newborn, and the progression of the disease in women with CF?

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Why is this important?

Today, most of the people with CF reach late adulthood and many people with CF chose to have children. In the literature, most of the studies done on pregnancy in women with CF are observational or compare women with CF with mild disease to patients without CF. In our study, we decided to compare mild to moderate-severe disease. We ascertain this will result in data that we can share, allowing individuals to make informed decisions using disease severity to guide choices around pregnancy.

What did you do?

We collected data on women with CF that had been pregnant between 1986-2018 from ten CF centres worldwide. Disease severity [mild or moderate-severe (mod-sev)] was defined according to lung function (forced expiratory volume % predicted in 1 second (FEV₁)) and body mass index (BMI) as well as functioning of the pancreas (pancreatic insufficiency (PI)) and *Pseudomonas aeruginosa* (PA) infection. Three time periods were compared, 12 months prior to conception, the pregnancy itself and the 12 months thereafter. Additionally, disease progression, pregnancy, delivery, and impact on health of the newborns (outcome) were examined and compared between the different groups.

What did you find?

Women with advanced disease had more CF-related complications during and after pregnancy and had increased preterm newborns. However, FEV₁ and BMI decline were no different between the mild and mod-sev groups.

A more rapid decline in FEV₁ was observed during pregnancy in PI and PA infected patients, though stabilizing thereafter.

When analysed according to disease severity, individuals with mod-sev disease had more flare-up of infections (pulmonary exacerbations (PEX)) during all three time periods, however, analysis over time showed no meaningful differences in the trends.

Women with chronic PA infection required fertility assistance more often and PI was associated with increased risk for infants with lower birthweight according to the duration of the pregnancy.

What does this mean and reasons for caution?

We conclude that CF disease severity defined by FEV₁, BMI, PA status and pancreatic status, has an impact on the outcomes of the pregnancy, delivery, and the newborn's status. Yet, the severity of the disease prior to the pregnancy does not seem to affect the annual decline in FEV₁, BMI or the number of PEX, and therefore, shows no impact on disease progression after

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pregnancy. Consequently, according to our study, we can assume that pregnancies in women with CF are in general well tolerated even in women with severe disease and no significant worsening of illness is expected during or after pregnancy.

What's next?

In Israel, on average every woman is getting 3.1 children (fertility rate). In the Jewish religious society, it can go up to 6.9 children/woman; and also, in the Arabic-Muslim population, there is a high rate of children/women. Many of our patients choose to undergo multiple pregnancies as a part of their cultural and religious lives. We are currently working on a study design to examine the effects of multiple pregnancies in women with CF.

Images: Three women with CF, with their children, with permission from them for publication.



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