Title: Pregnancy outcomes in women with cystic fibrosis and poor pulmonary function

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What was your research question? To investigate how poor pulmonary function (forced expiratory volume [FEV₁] below 50%) before women with cystic fibrosis (CF) became pregnant influenced the outcome of their pregnancy and how their clinical status developed.

Why is this important? Improved survival and overall health in CF have led to an increased number of women with CF making childbearing decisions. A number of studies have shown that having CF does not affect maternal pregnancy survival and does not affect maternal lung function, nutrition, and exacerbation rate. We also recently reported that there was no increased risk of deterioration of maternal pulmonary and nutritional status in women with CF and pre-pregnancy diabetes. Since the published pregnancy outcome data are limited for women with CF with poor pulmonary function, it is important to obtain data on this point.

What did you do? Pregnancies in women without lung transplantation with a first delivery reported to the French CF registry between 2000 and 2012 were identified. Pregnancy outcomes and clinical trends (body mass index – BMI, and pulmonary function) over a 4-year follow-up, starting the
year before pregnancy until two years after pregnancy, were compared between women with poor pre-gestational pulmonary function, defined as FEV₁ ≤ 50%, and women with FEV₁ > 50%.

What did you find?
A total of 149 women had a first delivery and 36 (24.2%) of these had pre-gestational FEV₁ ≤ 50%. There were no differences in age or frequency of assisted conception between the two groups. There were more cesarean sections in women with FEV₁ ≤ 50% (43.7% vs. 21.1%). The frequency of preterm birth did not differ between the two groups, but median infant birthweight was lower in women with FEV₁ ≤ 50% (2705g; range: 650-3700 vs. 3044g; range: 1590-3860). Despite lower FEV₁ and BMI the year before pregnancy for women with poor pulmonary function, the decline in these parameters during the study period did not differ between the two groups.

What does this mean and reasons for caution?
Poor pre-gestational pulmonary function in women with CF was associated with a higher rate of cesarean section and had a clinical impact on fetal growth, but it was not associated with greater pulmonary and nutritional decline over the study period. A sensitive analysis of women with very poor pulmonary function was not realized due to the small sample size of the group with low pulmonary function as it would not have been relevant on a statistical point of view, but might also contribute to the absence of difference.

What’s next?
In the present study, none of the patients were treated with CFTR modulators owing to the timing of the study. Further work is required to assess the risks and benefits of CFTR modulators in this context.

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