# La gravidanza nella donna FC

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Pregnancy and delivery in a patient with cystic fibrosis of the pancreas. Siegel B, Siegel S.

Segnalazione della prima gravidanza portata a termine con successo in una donna con FC.

> In quegli anni la mediana di sopravvivenza era meno di 10 anni.

> > Obstet Gynecol 1960;16:438-40.

### Distribuzione per età (RIFC)



Problematiche emergenti nella vita delle giovani donne FC

Inserimento nel mondo del lavoro
Vita di coppia
Gravidanza

### Pregnancy in Cystic Fibrosis\*

#### **Fetal and Maternal Outcome**

Marita Gilljam, MD; † Maria Antoniou, MD; Janey Shin, MSc; Annie Dupuis, MSc; Mary Corey, PhD; and D. Elizabeth Tullis, MD, FCCP

Results: From 1963 to 1998, there were 92 pregnancies in 54 women. There were 11 miscarriages and 7 therapeutic abortions. Forty-nine women gave birth to 74 children. The mean follow-up time was 11 6 8 years. One patient was lost to follow-up shortly after delivery, and one was lost after 12 years. The overall mortality rate was 19% (9 of 48 patients). Absence of Burkholderia cepacia (p<0.001), pancreatic sufficiency (p=0.01), and prepregnancy FEV1 > 50% predicted (p=0.03) were associated with better survival rates. When adjusted for the same parameters, pregnancy did not affect survival compared to the entire adult female CF population. The decline in FEV1 was comparable to that in the total CF population. Three women had diabetes mellitus, and seven developed gestational diabetes. There were six preterm infants and one neonatal death. CF was diagnosed in two children.

(CHEST 2000; 118:85-91)

The mean follow-up time was  $11 \pm 8$  years. One patient was lost to follow-up shortly after delivery', and one after 12 years. A double lung transplantation was carried out in 3 patients at 6, 10, and 13 years after the first completed pregnancy.

Five years after delivery, 26 women (90%) were alive and 3 women (10%) had died;

10 years after delivery, 19 women (79%) were still alive (Tables 2, 3). Death occurred in nine women at a median of 8 years (range, 3 to 30 years) after the first completed pregnancy. The causes of death were respiratory insufficiency and Cepacia syndrome.

Patient No.	Age at Diagnosis, yr	Age at Gestation, yr	Death Postpregnancy, yr	FEV <sub>1</sub> % Predicted Prepregnancy
1	0.5	32	3†	90
21	1.9	30	4	24
3	7.5	19	5	63
4§	2.7	25	6†	27
5	9.7	21	8	65
6§	12	29	13†	53
7	8.4	21	14	95
8†	24	22	25	34
9	57	28	30	NA

 $F = \Delta F508$ ; ? = unidentified mutation; + = positive, - = negative; NA = not available. † Cepacia syndrome.

‡ Medical termination of pregnancy. 3 years previously because of poor lung functions. § Lung transplantation later.

|| Another full-term pregnancy.

(CHEST 2000; 118:85-91)

#### The Effect of Pregnancy on Survival in Women With Cystic Fibrosis\*

Christopher H. Goss, MD, MS; Gordon D. Rubenfeld, MD, MS, FCCP; Kelly Otto, MS; and Moira L. Aitken, MD, FCCP

Measurements and results: Six hundred eighty of the 8,136 women in the cohort became pregnant. These 680 women were matched on an index year to 3,327 control women with CF. At the inception of entry into the cohort, women who reported pregnancy were more likely to have had a higher percentage of predicted FEV<sub>1</sub> (67.5% predicted vs 61.7% predicted, respectively; p < 0.001) and a higher weight (52.9 vs 46.4 kg, respectively; p < 0.001). Using Kaplan-Meier survival curves, the 10-year survival rate in pregnant women (77%; 95% confidence interval [CI], 71 to 82%) was higher than in those women who did not become pregnant (58%; 95% CI, 55 to 62%). A separate analysis, matching pregnant patients on FEV<sub>1</sub> percent predicted, age, *Pseudomonas aeruginosa* colonization, and pancreatic function, obtained similar results. Using Cox proportional hazard modeling to adjust for baseline age, FEV<sub>1</sub> percent predicted, weight, height, and pulmonary exacerbation rate per year, pregnancy was not associated with an increase risk of death. Pregnancy was not harmful in any subgroup including patients with FEV<sub>1</sub> < 40% of predicted or diabetes mellitus.

(CHEST 2003; 124:1460-1468)

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The pregnant cohort also had better survival among specific high-risk subgroups, as follows: FEV1, <40% predicted (p <0.001); and insulin-dependent diabetes mellitus (p<0.02)



*Conclusions:* Women with CF who became pregnant were initially healthier and had better 10-year survival rates than women with CF who did not become pregnant. After adjustment for the initial severity of illness, women who became pregnant did not have a significantly shortened survival. *(CHEST 2003; 124:1460-1468)* 

#### Cystic fibrosis and pregnancy. Report from French data (1980–1999)

Dominique Gillet<sup>a</sup>, Marc de Braekeleer<sup>a,b,c,d,\*</sup>, Gil Bellis<sup>d</sup>, Isabelle Durieu<sup>e</sup>, The participating centres to the French Cystic Fibrosis Registry

In our series, among 20 women with  $FEV_1 \leq 50\%$  before the first pregnancy, three (15%) died within the year following the end of pregnancy, while only one woman among 36 with  $FEV_1 > 50\%$  (3%) died during pregnancy. In fact, all women who died within the year after pregnancy had an  $FEV_1 < 50\%$ . So, in our point of view, pregnancy should be discouraged when patients reached this  $FEV_1$ level (which yet represents a bad health sign).

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### Parto prematuro

	Nati vivi	% parto prematuro
Edenborough e coll 2000 ( <i>UK)</i>	48	46%
Gilljam e coll 2000 ( <i>Canada)</i>	74	8%
Gillet e coll 2002 ( <i>Francia)</i>	55	18,2%
Odegard e coll 2002 ( <i>Scandinavia</i> )	33	24%

## Gravidanza a rischio

FEV1 < 50%</li>
 BMI < 20</li>
 diabete