

Pregnancy outcomes in women with cystic fibrosis; a Tasmanian case series

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Background:

With advances in medical care there has been an increase in the rate of pregnancy, and the desire for pregnancy, in women with Cystic Fibrosis (CF). These pregnancies are associated with increased risks - the mortality rate in women with CF is significantly higher than the background population, and high rates of premature delivery are also reported. Management of these women poses a challenge for health care practitioners. We present a case series describing the obstetric and neonatal outcomes in a cohort of women with CF.

Findings:

We report seven pregnancies in five women with CF over a six-year period; six singleton pregnancies and one dichorionic diamniotic twin pregnancy. One pregnancy was terminated at 12 weeks’ gestation due to severe respiratory disease, with pre-pregnancy FEV1 <30%. All other pregnancies resulted in live babies. Spontaneous conception occurred in five pregnancies, whilst two pregnancies were the result of assisted reproductive technology (ART). Women who conceived via ART had better respiratory function (higher pre-pregnancy FEV1) and received pre-pregnancy counselling.

Four patients had pancreatic insufficiency, with either pre-existing CF related diabetes or gestational diabetes mellitus.

Vaginal delivery occurred in two cases (median gestation 37.3 weeks). Elective caesarean section was performed in three cases for deteriorating maternal respiratory function (median gestation 31.9 weeks). One emergency caesarean section was performed for preterm labour in a twin pregnancy (gestation 27.5 weeks). There were no significant postpartum complications reported, however a drop in FEV1 and decreased maternal compliance with ongoing treatment was noted in three of seven cases.

There were no cases of fetal growth restriction. Neonatal admissions were all related to prematurity.

Table 1: Details of cases

| | 1 | 2 | 3 | 4 | 5 | 6 | 7 |
|---------------------------|-------------|-------------|-------------|-------------|----------------|------------------|------------------|
| Type of pregnancy | Singleton | Singleton | Singleton | Singleton | DCDA twins | Singleton | Singleton |
| Conception | Spontaneous | Spontaneous | Spontaneous | Spontaneous | ART | ART | Spontaneous |
| Pre-pregnancy counselling | No | No | No | No | Yes | Yes | No |
| FEV1 pre pregnancy | 53% | 35% | 41% | 30% | 63% | 82% | 64% |
| FEV1 at delivery | 42% | 29% | 39% | 37% | 77% | 81% | 55% |
| FEV1 post partum | 23% | 23% | 40% | 34% | 64% | 84% | 49% |
| Pregnancy outcome | Live birth | Live birth | Live birth | TOP | Live birth | Live birth | Live birth |
| Onset of labour | Elective | Elective | Elective | N/A | Preterm labour | IOL | IOL |
| Mode of delivery | Elective CS | Elective CS | Elective CS | N/A | Emergency CS | Vaginal delivery | Vaginal delivery |
| Gestational age | 34+3 | 30+3 | 31+1 | 12+3 | 27+5 | 37+1 | 37.5 |
| Time in NICU/SCN | 37 days | 49 days | 44 days | N/A | 67 days | Nil | Nil |

Key: ART: Assisted Reproductive Technology, CS: Caesarean Section, DCDA: Dichorionic Diamniotic, FEV1: Forced Expiratory Volume in one second, TOP: Termination of Pregnancy

Conclusions:

The findings of our study support the current literature of high rates of prematurity and caesarean section in women with CF. Pre-pregnancy counselling is recommended to optimise obstetric and neonatal outcomes. Multidisciplinary care between obstetric and respiratory physicians is paramount and women should be advised of the increased risk in pregnancy, and possible decline in respiratory function if treatment regimens are not adhered to.