Title	Pregnancy
Version	1.1

1. Introduction

There has been a steady rise in pregnancies in women with Cystic Fibrosis over recent years, in part related to improved health with CFTR modulator treatment. It remains important to ensure optimal health prior to pregnancy and to monitor closely to ensure the best possible outcomes for mother and baby.

2. Pre-conception counselling

Many women with Cystic Fibrosis will be in very good health and there may be no concerns about proceeding with pregnancy. However, it is important to ensure that lung function and (if present) diabetes control are optimised as much as possible. Ideally FEV1 would be above 60% predicted and HbA1c less than 48mmol/mol. Nutrition should also be optimised, with dietitian support, and a target BMI of 22kg/m^2 is recommended for best outcomes. Ideally pre-conception consultation by a specialist in maternal-fetal medicine should take place in collaboration with the CF team [1].

Prospective parents with cystic fibrosis may also wish to consider genetic testing for their partner to understand the risk of their child being affected by cystic fibrosis themselves. This can be arranged via referral to the CF team or to the Regional Genetics Service in Birmingham.

3. Medication review

All currently prescribed and over-the-counter medications should be reviewed by the CF team for their safety in pregnancy and any risks that may be associated with stopping treatment. This includes CFTR modulators. Inhaled mucolytics and inhaled/nebulised antibiotics are safe to continue in pregnancy but choice of Oral and IV antibiotics for any exacerbation may need to be adjusted [1].

Folic acid should be commenced at a dose of 400mcg daily if there are no other complications and 5mg daily if there is also a history of diabetes. Vitamin levels should be monitored and replaced if necessary, but Vitamin A dosage should not exceed 10,000iu per day [2] (for comparison the standard adult dose of Paravit-CF (2 capsules daily) contains 8,000iu of Vitamin A).

4. Obstetric care

Antenatal clinic review should be arranged monthly for the first 2 trimesters and 2-weekly in the 3rd trimester, with foetal growth scans every 2-4 weeks from 26 weeks onwards.

Routine anaesthetic review should be arranged at 12-16 weeks to discuss their role in pregnancy management and subsequent follow up at 26 weeks to formulate a plan for labour and delivery. Screening for gestational diabetes should be carried out if not already known to have diabetes.

5. Delivery and breast-feeding

Timing of delivery will depend on maternal and foetal health. In most women vaginal delivery is possible. Epidural analgesia may be of benefit.

Breast-feeding is encouraged after consideration of ongoing medication exposure, including to CFTR modulators. It is important to optimise nutritional intake to meet the increased calorie demand that breast-feeding requires.

6. Care following delivery

It is important to continue with a regular treatment routine, including nebulised therapy and airway clearance. If a caesarean section has been necessary then sufficient pain relief will be required to allow airway clearance to be performed comfortably and reduce the risk of chest infection related to retained secretions. It can be difficult to balance time for CF treatment with the demands of a newborn baby but it is important to ensure CF treatment is prioritised to prevent CF health related problems. Regular attendance at CF clinics is also important to ensure optimum monitoring.

Contraception should be considered at this stage. The combined oral contraceptive pill can be started 3-6 weeks after delivery in consultation with the obstetric/midwife team but should be avoided if breast-feeding. The progesterone only pill can be used during breast-feeding. Any other contraceptive method may also be considered. Lumacaftor/Ivacaftor (Orkambi) is known to reduce the effectiveness of hormonal contraception, but other current CFTR modulators do not.

7. References

[1] Jain R, Kazmerski TM, Zuckerwise LC, West NE, et al. Pregnancy in cystic fibrosis: Review of the literature and expert recommendations. J Cyst Fibros 2022;21:387-395
[2] Edenborough FP, et al. Guidelines for the management of pregnancy in women with cystic fibrosis. J Cyst Fibros 2008;7:S2-S32