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Pregnancy Care for women with Cystic Fibrosis Guideline

Introduction and Aim Provide guidance on the management of women with cystic fibrosis in pregnancy, including pre conceptual advice, antenatal, intrapartum, and postnatal considerations.	
Objectives Care of women during pregnancy and childbirth with cystic fibrosis	
Scope This policy applied to all healthcare professionals in all locations including those with honorary contracts.	
Equality Health Impact Assessment	An Equality Health Impact Assessment (EHIA) has not been completed.
Documents to read alongside this procedure	
Approved by	

Accountable Executive or Clinical Board Director	
Author(s)	Allison Condell, Gareth Lock, Pina Amin, Jamie Duckers, Lindsay George, Charlotte Addy, Ian Ketchell, Dawn Lau

Disclaimer

If the review date of this document has passed, please ensure that the version you are using is the most up to date either by contacting the document author or the Governance directorate.

Summary of review/amendments

Version Number	Date of Review approved	Date published	Summary of Amendments

Contents:

1. Introduction and Aim

Cystic fibrosis is a genetic condition, inherited in an autosomal recessive fashion that affects the quality and quantity of fluids produced by the many glands of the body. The hallmark of the condition is increased viscosity (or thickness) of these essential fluids, that results in varying levels of organ dysfunction. The organs principally affected by the condition are:

Lungs: Airways get blocked by thicker secretions resulting in bronchiectasis- a form of airways disease and recurrent, sometimes atypical, lung infections
Pancreas: Blockage of digestive enzymes, those that the pancreas usually excretes following a meal, results in malabsorption of vitamins and minerals, with subsequent malnutrition. Damage to the pancreas can occur over time which commonly results in CF related diabetes in 30-40% of adults with CF.
Hepatobiliary: Bile produced tends to be more viscous also, resulting in higher rates of gallstones. Gastro-oesophageal reflux rates are also higher in the CF population.
Reproductive organs: In both men and women subfertility rates are higher due blockages in the effective transfer of sperm & eggs. Approximately 99% of men with CF have negative Semen analysis due to CBAVD. Historically women with CF less fertile due to a combination of factors but since the introduction of CFTR modulators there has been a tripling of pregnancies
Skin: Increased salt loss is common in the sweat of CF patients
Liver: Increased risk of liver disease and porto-systemic hypertension; may present with varices of oesophagus.

In addition to the above, there is the psychological element of chronic disease that must be anticipated, particularly during the prenatal period. Psychological support can be arranged via psychologists within the CF service and/or via perinatal mental health services.
Due to advances in medical therapies and allied healthcare management of such patients, life expectancy and fertility rates have improved markedly over the last few decades, with a much longer life expectancy. As planning a family is now become a safer option, obstetric services in the UK are likely to encounter patients who live with CF becoming pregnant and requiring MDT specialist care. This guideline has been developed in conjunction with the All-Wales Adult Cystic Fibrosis Centre at CAVUHB to outline some of the differences in the management of such patients.

2. Pre-conceptual counselling

Working within a multidisciplinary team within a specialised centre can improve the outcome for the woman and her baby with due attention towards pro-active management of multi-system needs and challenges posed by the physiological needs of pregnancy and birth.

CF can have adverse outcomes on the fetus and 25% of the babies born to CF mothers are premature.

Generic obstetric guidelines aim to provide guidance for the care of uncomplicated pregnancies, but do not cover all aspects regarding specific medical conditions. Patients can be misinformed of their risks if we do not consult the specific information relevant to their condition, and cystic fibrosis patients are one such group. Therefore, pre-conceptual counselling is an important area of care for these patients who require discussion regarding medication and optimisation of health before getting pregnant.

2.1 Contraception and genetic testing

- Discuss contraception options whilst optimising in pre-conceptual phase- parenteral / coil may be advisable due to hepatobiliary disease and malabsorption
- Discuss fertility and partner testing / pre-implantation genetic diagnoses.

2.2 Maternal complications in pregnancy

- Increased risk of developing gestational diabetes in pregnancy or worsening of existing diabetes/difficulty controlling blood sugars.
- Susceptibility and severity of chest infections and the range of Abx that are safe to choose from in pregnancy (limited compared to non-pregnant population)
- Increased risk of intrahepatic cholestasis of pregnancy (Odds ratio 5x higher)- screen if symptomatic
- Increased risk of GORD- consider PPI in early pregnancy for hyperemesis

2.3 Fetal complications in pregnancy

- Increased risk of IUGR- will need growth scan assessments 28,32,36,39

2.4 Optimisation of maternal health to improve pregnancy outcomes

- Nutritional assessment- BMI and dietician involvement

- Lung function testing (3 monthly/home spirometry) {FEV1 <50% caution regarding pregnancy safety and outcomes}
- Cardiac function- may need Echo if FEV1 <50%, or signs of respiratory failure

- Some CF patients have liver disease- if portal HTN present, these patients may need endoscopy/discussion with gastro and regular LFTs
- Even in the absence of known liver disease, serial testing of LFTs is required for patients taking Kaftrio (Raised ALP/ALT is seen in deterioration/compare with previous results)
- Continue:
    - Regular physio (airway clearance and inhalation therapies, physical activity)
    - Pancreatic enzymes
    - Vaccination schedule
    - Discussion with CF team regarding when to stop Vit A supplement
- Offering to test baseline FBC, U&E, LFT, Hba1c

2.5 Medication for cystic fibrosis in pregnancy

- Medication review – optimising health whilst informing patient of known risks to medications particularly Kaftrio. See appendix 1 and 2
- Folic Acid 5mg once daily should be started as soon as contraception is stopped. Current advice is that folic acid should be initiated 3 months prior to planned conception.

3. Antenatal Care:

Specific antenatal considerations to discuss with the patient at booking (<10 weeks):

- Increased risk of IUGR- will need growth scan assessments 28,32,36,39
- Increased risk of intrahepatic cholestasis of pregnancy (Odds ratio 5x higher)
- Increased risk of GORD- consider PPI in early pregnancy for hyperemesis
- Increased risk of constipation and distal intestinal obstruction syndrome (a form of adult meconium ileus)- use of regular laxatives (in particular if patient taking iron)

It is imperative that the patient is encouraged to engage with obstetric and CF services throughout her pregnancy to aim for the best possible outcome.

At every appointment, the patient needs to be weighed, and saturations taken at each appointment.

Ensure vaccinations are up to date- Flu and COVID yearly.

Consultant review of VTE prophylaxis is required, taking into consideration disease activity, any long-term PORT lines, and obstetric risk factors for VTE.

11-13 weeks:

- As per routine care, a full antenatal booking assessment including routine bloods should be performed (FBC, U&E, LFT, ferritin). Include HbA1c for diabetes screening.
- Importance of chest physio from early pregnancy- pregnancy makes effective chest physio more challenging, patients may have to increase frequency of appointments
- Referral to specialist ANC in UHL at 16 weeks.
- Offer Aspirin as standard but hold if active haemoptysis. Dosage: Weight <70kg = 75mg, ≥70kg= 150mg. Stop aspirin temporarily if active haemoptysis and recheck platelets.

16 weeks:

- Initial consultant appt should be performed around 16 weeks of gestation to formulate individualise plan for antenatal care.

- Book the patient for serial growth scans 28, 32, 36 & 39 weeks
- Book anaesthetic assessment for 30/40

- Screen partner if not already screened
- MDT with CF team if obstetric concerns for specific cases. Email if not urgent, speak to team directly if more urgent. Input may be needed regarding dietician and physio appts.
    - Consider referral for antenatal anaesthetic review
- Commence Calcium and Vit D supplementation if not already on it from pre-conception (as per local guidelines).
- Early GTT testing for CF patients.

18-21 weeks:

- Routine Anomaly scan

From 28 weeks:

- Regular growth scans
- Anaesthetic assessment from 30-32 weeks.
- Weight and saturations, BP
- Screen for UTI, haematuria (renal stones more common), itching from third trimester (ICP)
- PNMH involvement- referral as required for appropriate team: CF psychologist/Obs Crisis team
- MoD discussion: Discussion with patient- liaise with CF and obs team.

Caring for a medically sick patient during antenatal period:

- If medically unwell- UHL admission to CF unit if <32 weeks gestation (if no signs of labour or obvious obstetric issues, for example bleeding or pre-eclampsia), with support from Obstetric team as required. Patients need to be cared for alike a normal UHW outlier (daily review with obs team attending from UHL antenatal unit). The CF unit should have a low threshold for contacting the obstetric team on call if they are admitting an unwell patient with CF.
- If patient is ≥32 weeks, for admission to UHW maternity unit with support from CF team, due to the need for regular fetal surveillance/higher risk of labour.
- Consultant to consultant discussion regarding place of care and management plan between obstetric team and CF team may be required for individual cases.

4. Induction of Labour and Intrapartum Care:

- If required, IOL in cystic fibrosis needs to be planned and discussed with the patient's named consultant.
- Women with CF can have a spontaneous vaginal birth if their lung function has been predicted to be at an appropriate range as has been agreed during the antenatal period.
- Up to 40% of CF patients deliver vaginally.
- When in established labour the woman should be transferred to the high dependency room on the delivery suite for continuous maternal and fetal monitoring. Consider 6 hourly U&Es in labour may be required if baseline abnormal.
- Adequate early analgesia in the form of low dose epidural analgesia during labour reduces cardiovascular and respiratory workload associated with labour.
- Allow prolonged passive stage of up to two hours to allow descent of head.
- It may be judicious to shorten the active second stage of labour to prevent prolonged Valsalva manoeuvre using forceps or vacuum as per clinical need. This should have been addressed with the patient in clinic in the antenatal period, but if not, clinical assessment should guide management.
- Women should be at the centre of discussion regarding MoD. Planned caesarean might be required in a proportion of women for example due to poor lung function that precludes vaginal birth. Liaise with the cystic fibrosis team (contact details at the end of guideline) with regards to the post operative care. Liaise with anaesthetist, core midwife and neonatal team to plan the C/S.
- Combined spinal epidural, epidural analgesia or general anaesthesia can be used for caesarean as dictated by the patient's CF status and anaesthetic input.
- Consider the need for antenatal corticosteroids and magnesium sulphate if preterm birth is planned. Women with diabetes will need variable rate intravenous insulin infusion (VRIII) as per protocol.
- Management of diabetes
    - VRII CANNOT BE PUT THROUGH PORTACATH/PICC LINE. Early liaison/low threshold with anaesthetics team for vascular access.
    - For women with diabetes / gestational diabetes who are having a planned caesarean section, the need for a variable rate intravenous insulin infusion (VRIII) should be discussed on an individual basis – most have sufficient endogenous insulin to cover background requirements when nil by mouth. Be aware of insulin sensitivity and close monitoring of BMs on insulin sliding scale. Discussion with diabetes team may be required early to control any swings in BMs (Joint British society for Perioperative care diabetes guideline, page 24-25)
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        - https://cpoc.org.uk/sites/cpoc/files/documents/2024-03/CPOC-DiabetesGuideline2023.pdf
    - Women having vaginal birth who have required insulin during pregnancy are likely to require a VRIII.
    - Women whose diabetes is managed by insulin pump could continue to use this during labour/C section at discretion of diabetes team and the wishes of individual.
    - If known pancreatic insufficient and on replacement enzymes even if NBM, patient would need to take an enzyme every 4 hours. Also, if on kaftrio and chosen to continue would ideally continue this medication even if NBM.

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5. Postnatal Care:

a. Postpartum care should be delivered in a high dependency room of the delivery suite for 24 hours.
b. The woman can be transferred to a side room with the baby on the postnatal ward to enable rest and minimise infection exposure. Adequate rest is necessary for optimal maternal recuperation and care of the newborn.
c. Extra help and support are needed for mothers with CF who have a new-born. This should be coordinated between the CF team, obstetricians, and midwives.
d. In many instances the women may need to continue IV antibiotics post-birth to clear infections.
    i. Management of PORT access. **Under no circumstances should the ports be accessed without prior training as it is essential the system is accessed correctly using the specialist type needle and flushed according to protocol**- routine venous access should be used, if unable to gain venous access then further support/training needs to be sought from the Cystic Fibrosis Clinical Nurse Specialists (CFCNS) based at UHL to liaise with nursing/midwifery staff at UHW. Consideration for patients to self-administer if trained.
    ii. Decision to transfer to oral antibiotics to be made in conjunction with CF team.
    iii. If required, newborn policy exists on CF unit if transfer is required.
e. Early mobilisation and assisted physiotherapy is critical in the postnatal period to help in clearance with clearance of the airways and improved ventilation. Within the first few hours of birth support with active chest clearance, inhaled therapies, and mobilisation in conjunction with adequate analgesia should be ensured.
f. Consultant review of VTE prophylaxis is required, taking into consideration disease activity, any long-term PORT lines, and obstetric risk factors for VTE.
g. Adequate postpartum analgesia helps with physiotherapy and early mobilisation.
h. If breast feeding is considered adequate nutritional supplementation of the mother should be ensured with additional intake of 500 kcal/day with addition of vitamin D, Calcium, and optimal hydration.
i. If mother is continuing Kaftrio, repeat LFTs for infant every 12 weeks for the duration of breast feeding.
j. Any medications omitted during pregnancy can be recommenced if not contraindicated should the mother consider breast feeding (Appendix 1 &2).
    i. Currently Kaftrio has insufficient evidence to support safe use in BF patients, so patients are advised to avoid. Discussion with patients antenatally revisited.
k. Appropriate initiation of contraception should be considered as soon as possible, ideally long-acting reversible contraception.

l. At discharge women with CF should have routine postnatal care with the community midwives, health visitors and other services and support systems available to new mothers.

6. Appendix 1:Drug use in pregnant and breastfeeding women with Cystic Fibrosis

Drugs	Recommendation in pregnancy	Recommendation for breastfeeding
Pancreatic enzymes	No risk is expected since no absorption by the mother.	Compatible. Poor systemic absorption.
Antibiotics		
Penicillins (+/- beta-lactamase inhibitors)	No risk was demonstrated. Use as first-line treatment.	Compatible. Some trace in milk may alter the gastrointestinal flora of the breastfed neonate.
Cephalosporins	No risk was demonstrated. Use as first-line treatment.	Compatible. Excreted in low concentrations in milk and may alter the gastrointestinal flora of the breastfed neonate.
Macrolides		
Erythromycin	No risk was demonstrated. Preferred drug in this class.	Possibly compatible. Very early exposure to erythromycin is associated with hypertrophic pyloric stenosis in infants. Low RID 1.4-1.7% [11]
Azithromycin	Probably no risk. Erythromycin should be preferred.	Probably compatible. RID is 3.9 % [11]
Carbapenems	Risks not well established. Limited data in human. Preferred drugs are either penicillin, cephalosporin, or erythromycin.	Poor oral bioavailability – unlikely to be absorbed.

Drugs	Recommendation in pregnancy	Recommendation for breastfeeding

Drugs	Recommendation in pregnancy	Recommendation for breastfeeding
CFTR Modulators*	Not recommended. Risks not established. Limited data in human.	Not recommended. Lumacaftor and ivacaftor detectable in milk and at low levels in plasma of breastfed infant [15].
Aminoglycosides	Risk of fetalnephro- and ototoxicity. Should be used only in case of life-threatening infections. Inhaled route can be used as low systemic absorption.	May alter the gastrointestinal flora of the breastfed neonate. For intravenous administration, avoid breast feeding within the 2 hours following injection. Inhaled route is safe for the breastfed infant.
Quinolones	Not recommended during pregnancy. If necessary, use the most documented drug of the class: ciprofloxacin.	Not recommended. High concentrations found in milk. RID > 10%.
Trimethoprim/ Sulfamethoxazole	Not recommended. Trimethoprim associated with neural defects when used in first trimester. Sulfonamide use in third trimester is associated with an increase in bilirubin in the neonate especially preterm.	Compatible in term neonates but avoid if G6PD deficiency. RID range from 4% to 9%.
Colistin	Risks not well established. Limited data in human. Intravenous route should be avoided. Inhaled route likely to be safe.	Possibly compatible with inhaled route.
Mucolytics		
rhDNase	Probably safe. Inhaled route associated with low systemic absorption.	Compatible since this large protein molecule is unlikely to be absorbed.
Hypertonic saline	Probably safe. Inhaled route associated with low systemic absorption.	Compatible. Poor systemic absorption.

- For more information see appendix 2: Kaftrio, pregnancy and breastfeeding: a guide for patients

7. Appendix 2: Kaftrio, pregnancy and breastfeeding: a guide for patients

Pregnancy and newborns:

Since the introduction of Kaftrio and Kalydeco, we have seen a significant increase in the number of women with CF becoming pregnant. We believe this is because of the effect Kaftrio has on the cervical mucous, making it thinner and easier to penetrate, and so this allows sperm to get through more easily. Along with this, the increased absorption of nutrients and vitamins in the gut and improved respiratory health in general means the body's cells are healthier and so more able to support a pregnancy. This is obviously exciting but has led to concerns regarding the safety of Kaftrio for the unborn baby. As things currently stand, the manufacturer (Vertex) has not tested this medication in pregnancy and so advice to avoid continuing to take it when a patient becomes pregnant. However, this medication has been used more extensively in the US and based on reports, there does not appear to be a major risk to continuing on the drug. We DO NOT know of any long-term risks to the baby at this time as Kaftrio has not been tested in this way before.

There is a theoretical risk of developing cataracts in young children on these medications; therefore, we do advise that any baby born to a mum on a CFTR modulatoris referred for eye screening by an ophthalmologist to ensure there is no evidence of cataract development. The risk of this is quite low though.

Kaftrio is passed through the placenta and cord to baby in the womb. As you know, all our patients who start on Kaftrio have liver function tests regularly. As the drug passes through the cord, we recommend that any baby born to a mum on a CFTR modulator should have liver function tests at

approximately 7 days after birth to ensure these are normal. There have only been a couple of cases of abnormal liver tests documented so the risk remains low.

We obviously want mum to be well and often our patients are well established on their Kaftrio. We know that suddenly stopping this medication can mean our patient's health deteriorates, which we obviously do not want in pregnancy. If you are aware of the small risk of cataracts and deranged liver tests to baby and that there are no known risks for mum (but usually a lot of benefits), then your CF team would support you continuing your Kaftrio. It is however YOUR decision.

One of the tests performed on newborns is the Newborn Bloodspot Screening test. This screening test is offered to all babies between day 4-6 of life (counting day of birth as 0) and screens for 9 rare conditions, including Cystic Fibrosis.

If the mother has been taking Kaftrio during pregnancy, this can pass through to the baby, potentially leading to a false negative result, even if the baby has CF.

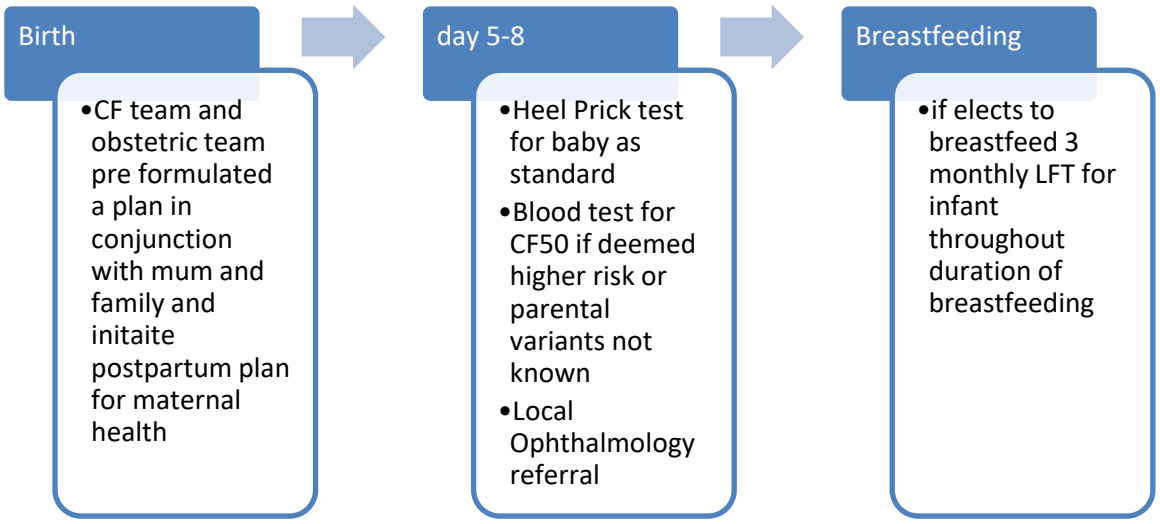
Ideally, we will already understand the likelihood of CF, as we would have offered genetic counselling to both parents before conception. If the risk is low, no further action is needed unless the baby develops symptoms later, in which case additional testing may be arranged.

If we are unsure about the parents' genetic information or if the risk of CF is higher, we will arrange for a blood test to check for a CF-related gene mutation. This blood test will be conducted around day 7, alongside other liver function tests. Results from the genetic test typically take about two weeks.

Breastfeeding:

There has been no formal research as to whether breastfeeding is safe for babies born to mum's who are taking Kaftrio. From initial studies, kaftrio is passed through breastmilk and so are passed to baby. As we said before, when any patient starts Kaftrio they must have regular blood tests for their liver function. This would mean that if you were to decide to breastfeed, then baby would need to have blood tests at least every 3 months or more often for the full duration of breastfeeding. We are generally recommending that mum's therefore do not breastfeed, but if this is something you really want to do, then please don't be afraid to discuss it further with your CF and obstetric team antenatally, or discuss with your midwife on the post-natal ward before discharge- that way we can ensure that baby is referred to their local paediatric team for this monitoring and that no harm comes to baby from the Kaftrio. There have been a few cases of abnormal blood tests in babies who have been breastfed in the US but there may have been other reasons for this.

Timeline if mum continued ETI through pregnancy



8. Contact details:

CF UHL Ward Manager: 25388 (Call for details for on call CF consultant)

CF Lead Specialist nurse UHL: 26488 (blp 6488)

CF Pharmacists: 25933

CF Dietetics: 26245

CF Physiotherapist: 26256 (blp 4989)/25913/07875565701

CF ward UHL (Doctor Office): 25028

CF FY1 ward doctor: (blp 4450)

CF F2 Doctor: (blp 4965)

CF UHL reception: 25599

UHL ANC: 02920726445/02920726103

UHW Labour ward: 42679/42686/41556

UHW Maternity triage unit: 44658/42635

UHW ward North (IOL suite): 46185

UHW ward East: 44436

UHW ward West: 46184

UHW Physio team (Bleep)

- Ward B7 Resp team: 0056
- Medical Resp Physio: 5878
- ITU/weekend Resp physio: 5295

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