

Policy

Clinical Guideline

Cystic Fibrosis in Pregnancy

Policy developed by: SA Maternal & Neonatal Clinical Network

Approved SA Health Safety & Quality Strategic Governance Committee on:

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Next review due: 30 June 2018

Summary Clinical practice guideline on cystic fibrosis in pregnancy.

Keywords Cystic fibrosis, CF, cystic fibrosis transmembrane conductance regulator, CFTR, cystic bronchiectasis, bronchopulmonary disease, forced expiratory volume, FEV1, non-invasive ventilation, Cystic Fibrosis in Pregnancy clinical guideline

Policy history Is this a new policy? **N**
Does this policy amend or update an existing policy? **Y v2.0**
Does this policy replace an existing policy? **N**

Applies to All Health Networks
CALHN, SALHN, NALHN, CHSALHN, WCHN, SAAS

Staff impact All Staff, Management, Admin, Students, Volunteers
All Clinical, Medical, Nursing, Allied Health, Emergency, Dental, Mental Health, Pathology

PDS reference CG214

Version control and change history

Version	Date from	Date to	Amendment
1.0	30 Jul 2007	18 Jan 2011	Original version
2.0	18 Jan 2011	24 Jun 2015	Reviewed
3.0	24 Jun 2015	Current	



South Australian Perinatal Practice Guidelines

cystic fibrosis in pregnancy

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Note

This guideline provides advice of a general nature. This statewide guideline has been prepared to promote and facilitate standardisation and consistency of practice, using a multidisciplinary approach. The guideline is based on a review of published evidence and expert opinion.

Information in this statewide guideline is current at the time of publication.

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Health practitioners in the South Australian public health sector are expected to review specific details of each patient and professionally assess the applicability of the relevant guideline to that clinical situation.

If for good clinical reasons, a decision is made to depart from the guideline, the responsible clinician must document in the patient's medical record, the decision made, by whom, and detailed reasons for the departure from the guideline.

This statewide guideline does not address all the elements of clinical practice and assumes that the individual clinicians are responsible for discussing care with consumers in an environment that is culturally appropriate and which enables respectful confidential discussion. This includes:

- The use of interpreter services where necessary,
- Advising consumers of their choice and ensuring informed consent is obtained,
- Providing care within scope of practice, meeting all legislative requirements and maintaining standards of professional conduct, and
- Documenting all care in accordance with mandatory and local requirements

Explanation of the aboriginal artwork:

The aboriginal artwork used symbolises the connection to country and the circle shape shows the strong relationships amongst families and the aboriginal culture. The horse shoe shape design shown in front of the generic statement symbolises a woman and those enclosing a smaller horse shoe shape depicts a pregnant women. The smaller horse shoe shape in this instance represents the unborn child. The artwork shown before the specific statements within the document symbolises a footprint and demonstrates the need to move forward together in unison.



Australian Aboriginal Culture is the oldest living culture in the world yet we experience the worst health outcomes in comparison. Our Aboriginal women are 2-5 times more likely to die in childbirth and our babies are 2-3 times more likely to be low birth weight. Despite these unacceptable statistics the birth of an Aboriginal baby is an important Cultural event and diverse protocols during the birthing journey may apply.

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Cystic fibrosis (CF)

- > Found primarily in Caucasian populations, CF is inherited as an autosomal recessive trait, caused by cystic fibrosis gene defect leading to dysfunction of the cystic fibrosis transmembrane conductance regulator (CFTR) protein¹
- > Approximately 1 in 25 people are carriers of the CF gene. Carriers of the CF gene do not have any symptoms of the condition
- > There are currently 300 people with CF in South Australia, with approximately 7 newborn babies diagnosed with CF each year
- > Mean survival of individuals with cystic fibrosis is now into the late thirties due to better disease management and treatment advances
- > CF is a recessive disorder, i.e. resulting from a mutated CFTR gene being inherited from each parent. When each of the parents has a CF mutation there is a 25% chance that the newborn will have CF, a 50% chance that the newborn will be a carrier, and a 25% chance of the newborn being a non-carrier

Introduction

- > Progressive chronic bronchopulmonary disease is the major cause of morbidity and mortality in cystic fibrosis (CF)
- > Major manifestations of cystic fibrosis include:
 - > Cystic bronchiectasis
 - > Pancreatic deficiency
 - > CF related diabetes
 - > CF related liver disease
 - > Small intestinal obstruction
 - > Sinus disease¹
- > Women with cystic fibrosis may have:
 - > Decreased fertility, generally related to decreased body mass index and unfavourable cervical mucus
- > Pre-pregnancy body weight and spirometry (especially forced expiratory volume in 1 second [FEV₁]) are useful predictors of maternal and neonatal outcomes²
- > Recurrent severe infection is a poor prognostic factor due to increased maternal mortality
- > Women with mild disease (FEV₁ > 80 % predicted) have been reported to tolerate pregnancy well, whilst those with advanced pulmonary disease (FEV₁ < 60 % predicted) may have more complications of both cystic fibrosis (infective exacerbations) and pregnancy (preeclampsia and gestational diabetes) but survival is similar to non-pregnant patients with cystic fibrosis³ (Geake et al. 2014). Compared to pregnant women without CF there is an increased rate of complications but the risks are small⁴. The most common adverse outcome for the fetus is preterm birth^{2,3}
- > Women with CF with pancreatic sufficiency generally maintain better spirometry and nutritional status than women with CF with pancreatic insufficiency

Pre-pregnancy counselling

- > Usually carried out by the CF Consultant and CF Multidisciplinary Team at the Royal Adelaide Hospital. Fertility clinics (Repromed and SA Fertility) and the South Australian Clinical Genetics Service at the Women's and Children's Hospital can also be involved in the counselling process
- > It is advisable for partners to be tested for carrier status and the risk of affected offspring calculated before pregnancy
 - > If the partner is not a carrier then the risk of the offspring having CF is low (less than 0.5 %)
 - > If the partner is a carrier, then the risk of the offspring having CF is 50 %. In this situation referral to the South Australian Clinical Genetics Service for counselling regarding reproductive options should be offered

Reproductive options if the partner is a carrier include:

- > An acceptance of the 50 % risk to offspring, with testing of the baby for CF after birth to allow treatment if affected
 - > Prenatal testing by CVS at 11 weeks of pregnancy with the option of termination of pregnancy if the fetus is shown to be affected
 - > Preimplantation Genetic Diagnosis, in which embryos conceived by IVF are tested and only those shown to be unaffected by CF chosen for transfer to try to establish a pregnancy
 - > Utilisation of a sperm or oocyte donor who is not a carrier for CF to reduce the risk of an affected child
 - > Choosing not to have biological children
- > Women with CF will be reviewed and monitored regularly by all members of the CF Multidisciplinary Team before and during pregnancy to optimise all aspects of health management. This may include assessments from the ;
- > Dietitian
 - > Physiotherapist
 - > Social Worker
 - > Clinical Psychologist
 - > Clinical Nurse
 - > Consultant
 - > Gastroenterologist
 - > Endocrinologist

Counselling:

- > Explain the need to continue treatments, including treatment for infective exacerbations. Attending required outpatient clinic appointments should also be emphasised. The SA Pharmacy Obstetric and Paediatric Medicines Information Service (located at the Women's and Children's Hospital) and the Adult Medicines Information Service (located at the Royal Adelaide Hospital) have collaborated on medicines information for women with Cystic Fibrosis in pregnancy
- > Discuss with the woman and her partner:
 - > The likely effect of pregnancy on the clinical course of CF (depending on the stage in her disease progression)
 - > Impact of CF and it's treatment on pregnancy outcomes (related to potential deterioration in spirometry during pregnancy), including risk of hospitalisation and preterm birth secondary to decline in respiratory function
 - > Issues around the burden of child rearing for someone with respiratory disease and the implication of premature death
- > Women with severe pulmonary disease ($FEV_1 < 60\%$) should be advised about the risks of pregnancy and coping with their disease post delivery
- > In the absence of reversible causes, delivery is the preferred treatment for respiratory failure

Factors associated with an increased maternal risk include:

- > Pulmonary hypertension
- > Cyanosis
- > Arterial hypoxemia (O_2 saturation $< 90\%$)
- > Moderate to severe lung disease ($FEV_1 < 60\%$ predicted)
- > Pre pregnancy evidence of poor nutritional status

Risks to the fetus

- > Preterm birth
- > Intrauterine growth restriction (uteroplacental insufficiency)
- > Cystic fibrosis

Antenatal care

- > Referral to a tertiary centre with Level VI facilities. Collaboration between this centre and the CF Multidisciplinary Team is vital
- > Joint initial assessment by a respiratory physician and an obstetrician experienced in dealing with problems of CF in pregnancy
- > If prenatal diagnosis of fetal CF is requested, consider chorionic villus sampling for early diagnosis and where requested, termination of an affected fetus

Particular attention to:

Dietary management

- > Assessed by the CF Dietitian
- > Maintenance of adequate nutrition – many women have pancreatic insufficiency and require enzyme supplements and a high caloric intake
- > Screen for Vitamin D deficiency and treat as required
- > Measure Vitamin A and E levels and supplement as required

Diabetes

- > Management of CF related diabetes – occurs in 20-30 % adults with CF and 15 % have impaired glucose tolerance
- > Oral Glucose Tolerance Test (OGTT) is recommended at initial booking visit, and repeated at, 20 to 24 weeks and at 30 to 34 weeks

Iron deficiency anaemia (IDA)

- > IDA is common in CF and oral supplementation is often indicated. Screen for anaemia at booking visit and again at 28 weeks
- > Encourage the woman to:
 - > Increase her dietary intake of iron
 - > Optimise absorption of iron by increasing intake of vitamin C and reducing foods that reduce bioavailability e.g. tannins (see Anaemia in pregnancy at www.sahealth.sa.gov.au/perinatal in the A to Z index for more information)

Respiratory management assessed by the CF physiotherapist

- > Baseline pulmonary function tests, such as FVC, FEV₁, lung volumes, pulse oximetry, and arterial blood gases as indicated. These values should not change appreciably in the early stage of pregnancy
- > Serial monitoring of values during gestation and address deterioration in pulmonary function
- > Early and adequate treatment of respiratory tract infections
- > Regular fetal growth monitoring (measure fundal height and serial ultrasound evaluations of fetal growth and amniotic fluid volume)
- > Anaesthetic referral and antenatal review

Anaesthetic review

- > Arrange early anaesthetic review to assess
 - > SpO₂
 - > Lung function
 - > Weight
 - > Diabetes
 - > If pulmonary hypertension is present

Other concerns include the presence of:

- > Severe respiratory disease and infection
- > Gastro-oesophageal reflux
- > In advanced lung disease the requirement for assisted ventilation should be considered
- > Adequate early analgesia in labour and flexible post-partum analgesia to permit physiotherapy and early mobilisation is advantageous

Intrapartum care

- > Management in consultation with obstetric physician
- > Aim for vaginal birth and reserve Caesarean section for obstetric indications
- > Epidural is preferred over general anaesthesia should operative delivery be necessary
- > Consider continuous external fetal monitoring as indicated
- > Continuous monitoring of maternal oxygen saturation using pulse oximetry and administer O₂ therapy as required. Non-invasive ventilation (NIV) may also be considered
- > Consider assisted birth if prolonged 2nd stage (predisposition to pneumothoraces)

Postpartum care

- > Recommend extra help and support with care of baby to prevent deterioration in maternal health
- > Encourage breastfeeding as long as adequate nutrition can be maintained to meet the increased energy demands
- > The commonly indicated CF drugs are safe in breastfeeding with the exception of co-trimoxazole (trimethoprim with sulfamethoxazole), which should be avoided in the first 4 weeks after birth or where the newborn baby is jaundiced because of the risk of kernicterus

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Safety of Cystic Fibrosis Medications in Pregnancy

NOTE: It is important to remember that **UNDERTREATMENT** may also be hazardous to a pregnant woman and her fetus

Agents which CAN BE USED in Pregnancy

ORAL MEDICATION

Calcitriol 0.25mcg (monitor neonate for hypocalcaemia)
Calcium
Cholecalciferol/Ergocalciferol
Omeprazole
Pancreatic Enzymes
Multivitamins
Ranitidine
Sodium chloride tabs
Ursodeoxycholic acid
Vitamin E (high doses must be reviewed)
Vitamin K1 (Phytomenadione)
Vitamin A There is sufficient evidence to recommend that vitamin A is safe in pregnant women up to a dose of 8000 IU / day. **VitABDECK®** contains 2500 IU / capsule and a daily dose of 2 caps per day is safe

INSULIN

INHALED MEDICATION

Beclomethasone
Budesonide or budesonide/eformoterol
Dornase Alfa
Fluticasone or fluticasone/salmeterol
Ipratropium
Mannitol
Salbutamol
Sodium Chloride
Terbutaline

ANTIBIOTICS – INHALED

Colistin
Tobramycin

ANTIBIOTICS – OTHER ROUTES

Azithromycin
Aztreonam
Cephalosporins – cefaclor, ceftazidime, cephalothin or cephalexin
Ciprofloxacin
Meropenem
Penicillins – amoxicillin, amoxicillin/clavulanate, dicloxacillin, flucloxacillin, phenoxymethylpenicillin, piperacillin/tazobactam or ticarcillin/potassium clavulanate
Tobramycin (human data suggests low risk however use IV only if inhalation therapy 300mg bd has failed)

Reference: Obstetric and Paediatric Medicines and Drug Information Services SA Pharmacy 2015

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Abbreviations

ADIPS	Australasian Diabetes in Pregnancy Society
BGL	Blood glucose level
CF	Cystic fibrosis
CFTR	Cystic fibrosis transmembrane conductance regulator protein
CVS	Chorionic villus sampling
et al.	And others
FEV ₁	Forced expiratory volume in 1 second
FVC	Forced vital capacity
e.g.	For example
GDM	Gestational diabetes mellitus
g	Gram(s)
Hb	Haemoglobin
HbA1c	Glycoslated haemoglobin
IVF	In vitro fertilisation
mg	Milligram(s)
NIV	Non-invasive ventilation
OGTT	Oral glucose tolerance test
RDS	Respiratory distress syndrome
SA	South Australia
WHO	World Health Organisation

Version control and change history

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