South Australian Perinatal Practice Guideline

Cystic Fibrosis in Pregnancy

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.

SA Health does not accept responsibility for the quality or accuracy of material on websites linked from this site and does not sponsor, approve, or endorse materials on such links.

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

Note: The words woman/women/mother/she/her have been used throughout this guideline as most pregnant and birthing people identify with their birth sex. However, for the purpose of this guideline, these terms include people who do not identify as women or mothers, including those with a non-binary identity. All clinicians should ask the pregnant person what their preferred term is and ensure this is communicated to the healthcare team.

"**Aboriginal and Torres Strait Islander recognition statement:** We use the term 'Aboriginal' to refer to people who identify as Aboriginal, Torres Strait Islander, or both Aboriginal and Torres Strait Islander. We do this because the people indigenous to South Australia are Aboriginal and we respect that many Aboriginal people prefer the term 'Aboriginal'. We also acknowledge and respect that many Aboriginal South Australians prefer to be known by their specific language group(s)."

Australian Aboriginal Culture is the oldest living culture in the world, yet Aboriginal people continue to experience the poorest health outcomes when compared to non-Aboriginal Australians. In South Australia, Aboriginal women are 2–5 times more likely to die in childbirth and their babies are 2–3 times more likely to be of low birth weight. The accumulative effects of stress, low socio-economic status, exposure to violence, historical trauma, culturally unsafe and discriminatory health services, and health systems are all major contributors to the disparities in Aboriginal maternal and birthing outcomes. Despite these unacceptable statistics, the birth of an Aboriginal baby is a celebration of life and an important cultural event bringing family together in celebration, obligation, and responsibility. The diversity between Aboriginal cultures, language and practices differ greatly and so it is imperative that perinatal services prepare to respectfully manage Aboriginal protocol and provide a culturally positive health care experience for Aboriginal people to ensure the best maternal, neonatal and child health outcomes.

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 horseshoe shape design shown in front of the generic statement symbolises a woman and those enclosing a smaller horseshoe shape depicts a pregnant woman. The smaller horseshoe 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.

Purpose and Scope of PPG

The guideline provides clinicians with recommendations for genetic counselling, management, and treatment for women with cystic fibrosis (CF) in the perinatal period.

Government of South Australia
SA Health

Table of Contents

Summary of Practice Recommendations

Couples should be counselled, and partners offered optimal testing for cystic fibrosis (CF) carrier status and the chance of having affected children discussed **before** pregnancy.

Couples identified to have a high chance of having a child affected by CF should be offered referral to the **Paediatric and Reproductive** branch of the **South Australian Clinical Genetics** Service **before** pregnancy to discuss reproductive options.

Pregnancy care should be provided in a level 5 or 6 maternity hospital involving Women's and Children's Hospital **Maternal Fetal Medicine** (WCH MFM), and the Royal Adelaide Hospital (RAH) CF multidisciplinary team.

Women with CF should be reviewed regularly at a frequency according to clinical status monthly in the first and second trimesters, and more frequently in the third trimester.

Spirometry and pulse oximetry should be performed at each clinical review.

Oral Glucose Tolerance Test (OGTT) is recommended as soon as pregnancy diagnosed if it has not been performed in the previous 6 months. If the result is negative, the test should be repeated at 12–16 weeks and again at 24 to 28 weeks' gestation

Anaesthetic review should occur early in pregnancy.

An intrapartum management plan should be developed in consultation with an obstetric physician.

Some women may be assessed as requiring adult intensive care facilities onsite for birth.

Continuous monitoring of maternal oxygen saturation using pulse oximetry during labour is recommended.

Vaginal birth is preferred over caesarean section to reduce post-birth pain which can impact chest clearance.

Neuraxial anaesthesia (epidural/spinal) is preferred over general anaesthesia should operative birth be necessary.

If carrier testing has shown an increased chance of having a CF affected baby and the couple opted to not test during pregnancy, the paediatrician should arrange genetic testing for Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) mutations in the baby to confirm if it is affected and requires ongoing management.

If the pregnant woman has taken CF modulator therapy during pregnancy, then her baby should have liver function checked at birth and screened for cataracts.

If breastfeeding mother is taking CFTR modulator therapy, then there should be ongoing monitoring of the infant including screening of liver function and eyes for the duration of the treatment.

Abbreviations

>	Greater than
≥	Greater than or equal to
<	Less than
≤	Less than or equal to
+/-	Plus or minus
CBAVD	Congenital bilateral absence of the vas deferens
CF	Cystic Fibrosis
CFTR	Cystic Fibrosis Transmembrane Conductance Regulator
CTG	Cardiotocography
CVS	Chorionic villus sampling
DIOS	Distal intestinal obstruction syndrome
EEG	Electroencephalogram
FEV_1	Forced expiratory volume in 1 second
FVC	Forced vital capacity
g	Gram(s)
ICU	Intensive care unit
IDA	Iron deficiency anaemia
IM	Intramuscular
IVF	In vitro fertilisation
IV	Intravenous
IUD	Intrauterine device
HbA1c	Haemoglobin A1c
kg	Kilogram(s)
mg	Milligram(s)
mL	Millilitre(s)
MFM	Maternal fetal medicine
Microg	Microgram(s)
NGS	Next-generation sequencing
NIV	Non-invasive ventilation
O_2	Oxygen
OCP	Oral contraceptive pill
OGTT	Oral glucose tolerance test
RAH	Royal Adelaide Hospital
SAPR	South Australian Pregnancy Record
USS	Ultrasound scan
WCH	Women's and Children's Hospital

Definitions

Shared decision making	Shared decision making involves discussion and collaboration between a consumer and their healthcare providers. It is about bringing together the consumer's values, goals, and preferences with the best available evidence about benefits, risks and uncertainties of screening, investigations, and treatment, to reach the most appropriate healthcare decisions for that person.
Spirometry	A pulmonary function test to measure lung function; including the amount and/or speed of air that can be inhaled and exhaled.

Background

Found primarily in Caucasian populations, cystic fibrosis (CF) is inherited as an autosomal recessive trait, caused by cystic fibrosis transmembrane conductance regulator protein (CFTR) gene mutations, leading to dysfunction of the CFTR protein.[1] 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. If the partner of a parent with CF is a carrier, there is a 50% chance the newborn will have CF. Approximately 1 in 25 people are carriers of mutations in the CF gene.[2] Carriers of the CF gene mutations do not have any symptoms of the condition.

There are currently 3,400 Australians with CF, with approximately 350 people with CF in South Australia.[2, 3] Approximately 7 newborn babies are diagnosed with CF each year. Predicted survival age of individuals with CF in Australia is 47 years, with new data reporting gains in life expectancy up to 61 years for those born after 2016.[2, 3] Progressive chronic bronchopulmonary disease is the major cause of morbidity and mortality in CF. Major manifestations of CF include:[1]

- cystic bronchiectasis
- pancreatic insufficiency
  - Women with CF with pancreatic sufficiency generally maintain better spirometry and nutritional status than women with CF with pancreatic insufficiency.
- CF related diabetes
- CF related liver disease
- small intestinal obstruction
- sinus disease.

In the past women with cystic fibrosis may have experienced decreased fertility, related to decreased body mass index and unfavourable cervical mucus[4, 5] and recurrent infection, which may have required oral and intravenous antibiotic therapy and close monitoring throughout pregnancy. However, with the availability of TRIKAFTA to approximately 90% of people living with CF, fertility has substantially increased due to the reversal of these complications.

Standards of Care

Eight standards of care have been developed by Cystic Fibrosis Australia and endorsed by the Thoracic Society of Australia and New Zealand to provide guidance on what is expected of healthcare providers and healthcare services to support people with CF:[5]

Standard 1	All young people with CF should be given timely, accurate information and counselling concerning fertility and pregnancy.
Standard 2	Preconception counselling should be offered to all women with CF and their partners. Pre-conception counselling should include CF genetic counselling for both the woman and reproductive partner, with discussion of the health implications of pregnancy
Standard 3	Assessment of fertility and appropriate referral to assisted reproduction services should be offered to women with CF who are planning parenthood.
Standard 4	Pre-pregnancy planning should include a detailed medical, nutritional, pharmacological, and mental health evaluation, assessment of glucose homeostasis and pre-pregnancy vaccinations. Access to psychosocial support should be provided as necessary.
Standard 5	Obstetric care should be provided as close as practical to a CF Centre. Care of woman during pregnancy should be provided by, or in regular consultation with, specialist CF multidisciplinary teams with experience in providing care for such pregnancies.
Standard 6	During pregnancy, women with CF should be reviewed frequently (minimum monthly) during the first and second trimesters and fortnightly during the third trimester by the CF Multidisciplinary Team to optimise pulmonary and nutritional health.
Standard 7	Clinical care during labour and delivery should be led by the obstetric team, with the CF team providing support. Choices about delivery should be made by the pregnant person and obstetrician.

Standard 8	Postpartum care should be jointly undertaken by the maternity and CF services with special emphasis on physiotherapy, nutritional and psychosocial support with adaptations for those delivering at other hospitals distant from the CF centre. The newborn screening status of the baby needs to be checked especially if exposed to CFTR modulators during pregnancy.

Pre-conception Counselling

Pre-conception counselling is usually carried out by the CF Consultant and CF Multidisciplinary Team at the Royal Adelaide Hospital (RAH). The Paediatric and Reproductive branch of the South Australian Clinical Genetics Service at the Women's and Children's Hospital (WCH), WCH Maternal Fetal Medicine (MFM), and genetics staff at private fertility clinics may also be involved in the counselling process. An important aspect of pre-conception counselling and planning for people with CF, is genetic testing.[4]

- Partners should be offered testing for CF carrier status and the chance of affected children calculated **before** pregnancy.[5]
  - The optimal method for the partner's carrier testing is next-generation sequencing (NGS) based CFTR gene sequencing (including deletion/duplication analysis) to ensure the maximal chance of detecting if a partner is a carrier.
  - This may be as a standalone test (ideally through SA Pathology) under Medicare or as part of a larger reproductive carrier screening panel. Screening for *women* planning pregnancy for CF status, spinal muscular atrophy status and Fragile X gene status ("three genes screen" is now a Medicare rebatable test (73451) available through a range of laboratories, including SA Pathology. The CFTR gene is also included in most expanded Reproductive Carrier Screening panels offered by private laboratories which offer screening for carrier status for many hundreds of conditions at a cost to the patient.

CAUTION is required in selecting a laboratory if the CFTR screening is or has been done as part of a "three gene screen" or larger reproductive carrier screening panel, as different laboratories utilise different methodologies for CFTR screening, many of which are suboptimal for this situation. (i.e., a panel of common variants rather than sequencing and non-reporting of the intron 8 poly T tract status).

- A specific request to include the intron 8 poly T tract status in the partner's testing result should be made, as this is **not** routinely reported in a carrier screening situation.
- If the partner is not identified to be a carrier, then the chance of a child being affected by CF is low (less than 0.1%), however, all children will be obligate carriers for CF.
- If the partner is a carrier, then there is a 50% (1 in 2) chance of a child being affected by CF and a 50% (1 in 2) chance a child will be a carrier of CF. Refer couple to the Paediatric and Reproductive branch of the South Australian Clinical Genetics Service (PRGU) at the WCH or a private IVF service offering genetic counselling services for counselling regarding reproductive options.
  - Reproductive options if the partner is a carrier include:
    - an acceptance of the 50% chance of an affected child with testing of the baby for CF after birth to allow treatment if affected.
    - prenatal testing by CVS at 11-12 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. This is accessible in SA through several Private IVF providers with partial Medicare coverage, but potential out of pocket cost to a couple.
    - utilisation of a sperm or oocyte donor who after appropriate screening has been found not to be a carrier for CF to reduce the risk of an affected child.
    - choosing not to have biological children +/- consider alternative options for parenting such as adoption or fostering.

- If a couple are both affected by CF then there is a 100% chance a child will be affected by CF.
  - Refer couple to the Paediatric and Reproductive branch of the South Australian Clinical Genetics Service at the WCH (PRGU) for counselling regarding reproductive options.
  - Following counselling regarding the full range of reproductive options the PRGU is able to refer couples on to Private IVF units offering Preimplantation genetic diagnosis and gamete donation or services providing further information regarding fostering/adoption, if these are pathways a couple express further interest in.
- 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:
  - Dietitian
  - Physiotherapist
  - Social Worker
  - Clinical Psychologist
  - Clinical Nurse
  - Consultant
  - Gastroenterologist
  - Endocrinologist

Aboriginal women should be referred to an Aboriginal Health worker, AMIC or Aboriginal Health Practitioner to ensure cultural advice and support in planning their care.

- Explain the need to continue treatments, including treatment for infective exacerbations. Attending required outpatient clinic appointments should also be emphasised.
- Discuss with the woman and her partner:
  - If on CF modulator therapy: The potential unknown risks to the developing fetus of CF modulatory therapy, as well as the growing evidence that there has so far been no significant increase in congenital abnormalities or premature birth for those who remain on therapy, versus the risk of discontinuation of modulator therapy to the mother according to the woman's clinical status both pre modulator therapy and currently. If the mother's clinical status is not a significant risk, then recommendation is to cease modulator therapy three months prior to ceasing contraception or commencing conception attempts
  - the likely effect of pregnancy on the clinical course of CF (depending on the stage in her disease progression and suitability of modulator therapy)
  - impact of CF and its treatment on pregnancy outcomes (related to potential deterioration in lung health during pregnancy), including risk of hospitalisation and preterm birth secondary to decline in respiratory function
  - adjustments to regular medications if there is a risk to the developing fetus
  - issues around the burden of child rearing for someone with respiratory disease and the implication of premature death according to mother's clinical state, stability and suitability for modulator therapy
  - women with severe pulmonary disease (FEV_1 < 60%) should be advised about the risks of pregnancy and coping with their disease post birth
  - in the absence of reversible causes, birth is the preferred treatment for respiratory failure or deterioration.

Aboriginal women should be offered an Aboriginal Health Care Worker or Aboriginal Cultural Support, when being consulted on decision relation to their care and treatment.

Factors Associated with an Increased Maternal Risk

- 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

Risk to the Fetus

- Preterm birth
- Intrauterine growth restriction (uteroplacental insufficiency)
- CF
- Side effects of medications

Antenatal Care

- Care should be provided in a level 5 or 6 maternity hospital involving WCH MFM, and the RAH CF multidisciplinary team (see *Maternal & Neonatal Services Standards* available at www.sahealth.sa.gov.au/perinatal).
  - Some women may be assessed as requiring adult intensive care facilities onsite for birth and this will affect choice of birthing hospital.
  - Some women with minimal disease may elect antenatal care closer to home or private practice, and individual management plans should be developed in conjunction with the proposed treating practitioner/team and WCH MFM and RAH CF services in this setting.

> ⚠ Most of the available evidence is from the pre- cystic fibrosis transmembrane conductance regulator (CFTR) modulators era. The following recommendations are now attenuated according to maternal clinical status, both pre-pregnancy and as the pregnancy progresses.

The general recommendation is to cease CFTR modulators pre pregnancy if safe to do so, however, if there are any concerns for maternal health throughout pregnancy where mother is not taking CFTR modulator and is eligible to, the first consideration is whether these should be re-introduced.

- Pre CFTR modulator therapy it is recommended that women with CF should be reviewed at least monthly in the first and second trimesters, increasing to every 2 weeks in the third trimester.[5, 6]
  - More frequent follow-up, if recommended, during pregnancy for women with more serious disease.[6]
- Overall health status at the time of conception can be indicative of obstetric and neonatal outcomes.[4, 5]
  - Pre-pregnancy body weight and spirometry (especially FEV_1, are useful predictors of maternal and neonatal outcomes.
- Women with mild disease (FEV_1 > 80% predicted) have been reported to tolerate pregnancy well, whilst those with advanced pulmonary disease (FEV_1 < 60% predicted) may have more complications of both CF (infective exacerbations) and pregnancy (preeclampsia and gestational diabetes), but survival is similar to non-pregnant women with cystic fibrosis.[4, 7] Compared to pregnant women without CF, there is an increased rate of complications but the risks remains small.[4] The most common adverse outcome for the fetus is preterm birth.[2,3]
- An obstetrician and obstetric physician should be involved in the woman's care and document the management plan in the SA Pregnancy Record (SAPR) and medical record.
  - Collaboration with the CF Multidisciplinary team is vital.

- If the woman and/or couple are known to be at increased risk of having a child with CF based on carrier screening of the partner, and prenatal diagnosis of fetal CF is requested, refer the woman to the Paediatric and Reproductive branch of the South Australian Clinical Genetics Service and WCH MFM.
- Consider chorionic villus sampling (CVS) for early diagnosis and where requested, termination of an affected fetus.
  - CVS is the preferred option undertaken between 11 and 13 weeks.
- Amniocentesis is an alternative undertaken from 15 weeks.
- The option of termination of pregnancy will be available if the fetus is shown to be affected by CF.

Priorities of Care

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.
- Measure Vitamin A, E and D levels and supplement as required.

For Aboriginal women in rural/remote locations speak with an Aboriginal Community Controlled Health Service, where applicable, to discuss a plan to ensure access to supplements is available.

Diabetes

- Gestational and pregestational diabetes is highly prevalent in women with CF, due to lower insulin secretion, increased insulin resistance and increased hepatic glucose production.[8]
- Oral Glucose Tolerance Test (OGTT) is recommended at:[8-10]
  - Pre-pregnancy (as soon as pregnancy identified if OGTT > 6 months prior)
  - 12–16 weeks' gestation
  - 24–28 weeks' gestation.
- If diabetes confirmed, target HbA1c < 6.5% at the time of conception and < 6% during pregnancy in combination with glucose measurements to confirm achieving appropriate glycaemic control.[11]

Iron Deficiency Anaemia (IDA)

- IDA is common in CF and oral supplementation is often indicated.
- Screen for anaemia at:
  - initial booking visit, and
  - 28 weeks gestation.
- 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 PPG* found in the A-to-Z index at www.sahealth.sa.gov.au/perinatal).

Consider IV iron as first line treatment for IDA for Aboriginal women and women in rural/remote communities where accessibility and availability of iron-rich foods and supplements may be limited.

Respiratory Management

- Joint initial assessment by a respiratory physician and an obstetrician experienced in dealing with problems of CF in pregnancy should be performed.
- Ongoing respiratory management by a CF physician, should include:
  - baseline spirometry and pulse oximetry
    - 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.

Anaesthetic Review

- Arrange early anaesthetic review to assess:
  - SpO_2
  - lung function
  - weight
  - diabetes
  - If pulmonary hypertension is present.
- Other concerns include the presence of:
  - severe respiratory disease and infection
  - gastro-oesophageal reflux
  - recurrent constipation and distal intestinal obstruction syndrome (DIOS)
  - mental health conditions
- 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.

Cystic Fibrosis Medications in Pregnancy

The safety of cystic fibrosis transmembrane conductance regulator (CFTR) modulators in pregnancy and lactation are being established.[12] 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 (see Table 1).

***Note**: It is important to remember that undertreatment may be hazardous to a pregnant woman and her fetus.*

Intrapartum Care

- Management plan should be developed in consultation with an obstetric physician.
- Aim for vaginal birth. Caesarean section should be reserved for obstetric indications.
- Consider continuous electronic fetal monitoring.
- Continuous monitoring of maternal oxygen saturation using pulse oximetry and administer O_2 therapy as required.[5]
  - Non-invasive ventilation (NIV) may also be considered.
- Consider assisted birth if prolonged 2nd stage (predisposition to pneumothoraces if lung disease present).
- Neuraxial anaesthesia (epidural/spinal) is preferred over general anaesthesia should operative birth be necessary.[5]
- Adequate early analgesia in labour and flexible post-partum analgesia to permit physiotherapy and early mobilisation is advantageous.

Table 1: Medications that can be given to women with cystic fibrosis in pregnancy.

Oral Medication	• Calcitriol 0.25 microg (monitor neonate for hypocalcaemia) • Calcium • Cholecalciferol/Ergocalciferol • Omeprazole • Pancreatic Enzymes • Multivitamins • Sodium chloride tablets • 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 for pregnant women up to a dose of 8000 international units/day. VitABDECK® contains 2500 international units/capsule and a daily dose of 2 caps per day is safe).
Insulin	• All
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 • Meropenem • Penicillins: amoxycillin, amoxycillin/clavulanate, dicloxacillin, flucloxacillin, phenoxymethylpenicillin, piperacillin/tazobactam or ticarcillin/potassium clavulanate • *Ciprofloxacin and IV Tobramycin are **not** recommended in pregnancy* •

Postpartum Care

- Recommend extra help and support with care of baby to prevent deterioration in maternal health.
- Encourage breastfeeding as long as adequate maternal nutrition can be maintained to meet the increased energy demands.

***Note**: Where indicated, consider liver function tests for the neonate where there is ongoing exposure to CFTR modulators through lactation.*[12]

- The commonly indicated CF drugs are safe in breastfeeding except for co-trimoxazole (trimethoprim with sulfamethoxazole), which should be avoided in the first 4 weeks after birth or where the newborn baby is jaundiced due to the risk of kernicterus.
- If carrier testing has shown an increased chance of having a CF affected baby and the couple opted to not test during pregnancy, the paediatrician should offer genetic testing for CFTR mutations in the baby to confirm if it affected and requires ongoing management.

Where possible, an Aboriginal Community Controlled Health Service should be contacted to discuss postpartum care when woman and family return to country. An Aboriginal health professional should be engaged to seek possibilities for timely and safe care, in addition to timely referral to a health service or primary carer.

Resources

Australian Government Pregnancy, Birth and Baby: (www.pregnancybirthbaby.org.au)
Pregnancy, Birth and Baby | Pregnancy Birth and Baby (pregnancybirthbaby.org.au)

Cystic Fibrosis Australia:
www.cysticfibrosis.org.au

Cystic Fibrosis South Australia:
https://www.cfsa.org.au/

Information for new Parents:
Consumer booklet: Information for parents with a baby diagnosed with cystic fibrosis

Medicines Information: (sahealthlibrary.sa.gov.au)
https://sahealthlibrary.sa.gov.au/friendly.php?s=SAPharmacy

Pathology Tests Explained: (https://pathologytestsexplained.org.au/)
Pathology Tests Explained

SA Health Pregnancy:
Pregnancy | SA Health

SAPPGs Web-based App:
Practice Guidelines (sahealth.sa.gov.au)

References

1. Bienvenu T, Lopez M, Girodon E. Molecular Diagnosis and Genetic Counseling of Cystic Fibrosis and Related Disorders: New Challenges. Genes (Basel). 2020;11(6).
2. Cystic Fibrosis South Australia. Living with Cystic Fibrosis: Cystic Fibrosis SA Inc; [Available from: https://www.cfsa.org.au/.
3. Australian Cystic Fibrosis Data Registry. 2023 Annual Report: Monash University; 2024. Available from: https://www.cysticfibrosis.org.au/wp-content/uploads/2024/07/ACFDR_2023_Annual-Report.pdf.
4. Jain R, Kazmerski TM, Zuckerwise LC, West NE, Montemayor K, Aitken ML, et al. Pregnancy in cystic fibrosis: Review of the literature and expert recommendations. J Cyst Fibros. 2022;21(3):387-95.
5. Douglas TA, Mulrennan S, Fitzgerald DA, Prentice B FK, Messer M, Bearcroft A, et al. Standards of Care for Cystic Fibrosis. Sydney: Cystic Fibrosis Australia; 2023. Available from: https://www.cysticfibrosis.org.au/wp-content/uploads/2024/07/TD_CFA_Standard-of-Care_Design_Refresh_2024.07.25.pdf.
6. Middleton PG, Gade EJ, Aguilera C, MacKillop L, Button BM, Coleman C, et al. ERS/TSANZ Task Force Statement on the management of reproduction and pregnancy in women with airways diseases. Eur Respir J. 2020;55(2).
7. Geake J, Tay G, Callaway L, Bell SC. Pregnancy and cystic fibrosis: Approach to contemporary management. Obstet Med. 2014;7(4):147-55.
8. Oxman R, Roe AH, Ullal J, Putman MS. Gestational and pregestational diabetes in pregnant women with cystic fibrosis. J Clin Transl Endocrinol. 2022;27:100289.
9. Khare S, Desimone M, Kasim N, Chan CL. Cystic fibrosis-related diabetes: Prevalence, screening, and diagnosis. Journal of Clinical & Translational Endocrinology. 2022;27:100290.
10. Moran A, Brunzell C, Cohen RC, Katz M, Marshall BC, Onady G, et al. Clinical Care Guidelines for Cystic Fibrosis–Related Diabetes: A position statement of the American Diabetes Association and a clinical practice guideline of the Cystic Fibrosis Foundation, endorsed by the Pediatric Endocrine Society. Diabetes Care. 2010;33(12):2697-708.
11. Montemayor K, Tullis E, Jain R, Taylor-Cousar JL. Management of pregnancy in cystic fibrosis. Breathe. 2022;18(2).
12. Jain R, Taylor-Cousar JL. Fertility, Pregnancy and Lactation Considerations for Women with CF in the CFTR Modulator Era. Journal of Personalized Medicine. 2021;11(5):418.

Acknowledgements

The South Australian Perinatal Practice Guidelines gratefully acknowledge the contribution of clinicians and other stakeholders who participated throughout the guideline development process particularly:

Write Group Lead

Jan Leibelt

Write Group Members

Dr Judith Morton
Belinda Nitschke

Other Major Contributors

Bianca Budin
Dr Danielle Crosby
Nicole Cullinan
Dr Gemma Hardi
Dr Michael McEvoy
Dr Linda McKendrick
Dr Anthia Rallis
Dr Amanda Poprzeczny

SAPPG Management Group Members

Dr Michael McEvoy (Chair)
Monica Diaz (SAPPG EO)
Dr Elizabeth Allen
Elise Bell
Dr Angela Brown
Marnie Campbell
John Coomblas
Dr Danielle Crosby
Imogen Downard John
Kate Greenlees
Luke Grzeskowiak
Kathryn Hansen
Dr Gemma Hardi
Dr Susie Keynes
Belinda Nitschke
Dr Angela Mathew
Dr Scott Morris
Dr Amanda Poprzeczny
Dr Anthia Rallis
Amy Rigano
Dr Shruti Tiwari
Sally Wadsworth
Allison Waldron

Suggested citation:

Leibelt J. Cystic Fibrosis in Pregnancy [Internet]. Version 4. Adelaide (AU): South Australian Perinatal Practice Guideline; SA Health, Government of South Australia; 2025. 14 p. Guideline No.: PPG214. Available from: http://www.sahealth.sa.gov.au/perinatal.

OFFICE USE ONLY

Document Ownership & History

Developed by: Maternal, Neonatal and Gynaecology Strategic Executive Leadership Committee
Contact: HealthCYWHSPerinatalProtocol@sa.gov.au
Approved by: Clinical Guidelines Domain Custodian
Next review due: 12/07/2030
CGSQ reference: PPG214
Guideline history: Is this a new perinatal practice guideline (V1)? **N**
Does this perinatal practice guideline amend or update an existing perinatal practice guideline? **Y**
If so, which version? **V3**
Does this perinatal practice guideline replace another perinatal practice guideline or policy with a different title? **N**
If so, which perinatal practice guideline or policy (title)?

Approval Date	Version	Who approved New/Revised Version	Reason for Change
12/07/2025	V4.0	Clinical Guidelines Domain Custodian	Formally reviewed in line with 1–5 yearly scheduled timeline for review. Updated resources.
24/06/2015	V3.0	SA Health Safety & Quality Strategic Governance Committee.	Formally reviewed in line with 1–5 yearly scheduled timeline for review.
18/01/2011	V2.0	SA Health Safety & Quality Strategic Governance Committee.	Reviewed.
30/07/2007	V1.0	SA Maternal and Neonatal Clinical Network	Original approved version.