

# Provision of contraception services and advice for women with cystic fibrosis

Sarah Gatiss, Diana Mansour, Simon Doe, Stephen Bourke

## Abstract

**Background and methodology** As the prognosis of patients with cystic fibrosis (CF) improves, issues of sexual health, fertility, pregnancy and contraception are increasingly important. In order to plan the provision of a contraception and sexual health service for women with CF we studied their sexual and reproductive history, their current usage of contraception, the sources and quality of advice they had received, and their particular needs using a confidential questionnaire sent to all women over 16 years of age attending a regional CF centre.

**Results** Of 55 women (mean age 29.7 years) surveyed, 42 (76%) responded. Thirty-three women (79%) were sexually active and 13 (31%) had experienced 19 pregnancies, five (26%) of which were unplanned. Only half of the women who responded were using contraception. No woman used female sterilisation, the progestogen implant, intrauterine system (IUS) or copper-

bearing intrauterine device (IUD) for contraception. Twenty-six (62%) women reported not having received contraceptive advice specific to CF and 24 (57%) said that they had not been warned about the potential interaction between broad-spectrum antibiotics and the combined pill.

**Discussion** Women with CF have a relatively high rate of unplanned pregnancy and do not receive optimal advice or use the full range of contraceptive methods. CF teams lack training in contraception and contraceptive services may not have a detailed knowledge of CF and its complications. New strategies are needed to focus the knowledge and skills of both teams in providing better services for women with CF.

**Keywords** contraception, cystic fibrosis, fertility, pregnancy, sexual health care

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## Introduction

Contraception and sexual health services aim to provide easy access to all methods of contraception. Particular difficulties may arise in providing specialist contraceptive advice to women with complex medical conditions such as cystic fibrosis (CF).

CF is an autosomal-recessive disease resulting from mutations of the CF gene, which encodes for a protein that regulates the chloride channel in the membrane of epithelial cells. Reduced chloride conduction results in viscid secretions and organ damage in the respiratory, gastrointestinal, pancreatic, hepatobiliary and reproductive tracts. In the UK about 1 in 2500 children are born with CF, 1 in 25 of the population is a carrier of the disease, and of the 7500 people living with CF 50% are aged over 16 years. The major cause of death is respiratory failure due to progressive lung infection and bronchiectasis. About 85% of patients have pancreatic insufficiency with malabsorption of fat. With increasing age patients may develop complications such as diabetes, osteoporosis, liver disease and gallstones. Patients receive complex treatment regimens including chest physiotherapy, nutritional supplements, pancreatic enzymes and continuous oral or inhaled antibiotics. They also receive frequent additional

### Key message points

- Most women with cystic fibrosis (CF) are fertile, but pregnancy should be planned so that the mother's health can be optimised, teratogenic drugs avoided and genetic counselling considered.
- Women with CF have a relatively high rate of unplanned pregnancy and do not receive optimal advice or use the full range of contraception methods.
- CF teams lack training in contraception and sexual health clinics may not have a detailed knowledge of CF.

courses of oral antibiotics or intravenous antibiotics administered via implanted venous access systems.

The median survival of patients has improved to about 30 years currently, but is projected to improve further to almost 50 years for a child born in 2000.<sup>1,2</sup> More than 1500 mutations of the CF gene have been identified and there is a wide range of disease severity with some patients having good overall health into their fourth decade or later. As the prognosis of patients with CF improves, the issues of sexual health, fertility, pregnancy and contraception are increasingly important.

The female genital tract in CF is anatomically normal but the cervical mucus is thickened and may act as a barrier to sperm. Some women with severe disease may have amenorrhoea or anovulatory menstrual cycles. However, most women with CF are fertile, with 75% of those who try to conceive becoming pregnant.<sup>3</sup> Ideally pregnancy should be planned so that the mother's health can be optimised, teratogenic drugs avoided, and genetic counselling considered. Pregnancy is inadvisable in those with severe lung disease and maternal deaths have occurred. Although the outcome of pregnancy in CF is generally good, pregnancy and motherhood place additional burdens on the mother's health. Women wishing to avoid pregnancy need specialist advice as some contraceptive methods may be contraindicated and efficacy may be reduced. They also need to be aware of all contraceptive choices, the effects

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that contraceptive hormones may have in relation to their disease, and the effect that pregnancy may have on their health.<sup>4-6</sup> Provision of contraceptive services to these women may be difficult as CF physicians lack training in all contraceptive options and family planning services may not have an adequate understanding of CF and its complications. In order to plan the provision of services we undertook a survey of women with CF, focusing on their knowledge of reproductive issues and their contraceptive requirements.<sup>3</sup>

## Methods

A confidential questionnaire was designed in a series of meetings between the CF team and the staff of the contraception and sexual service. It was then piloted in the CF clinic before being posted to all women attending the regional CF centre. The questionnaire was divided into sections addressing obstetric history, current relationships and future pregnancy plans; current and previous use of contraception; and knowledge and source of sexual health advice. The questionnaire was sent again to non-responders and at their next appointment clinic staff also asked non-responders if they would be happy to complete the questionnaire. The data were analysed using Microsoft Excel™.

## Results

Of the 55 women surveyed (mean age 29.7 years, range 16–51 years), 42 (76%) responded. Thirty-five women responded to the initial postal survey, two responded to a secondary 'reminder' questionnaire and five completed the questionnaire when they attended the CF clinic. Of the 13 non-responders, two were pregnant at the time of the survey and one had undergone a hysterectomy. No further information was obtained for the remaining 10 women.

Eleven (26%) women were single: eight (19%) had no partner, two were divorced and one was separated from her husband. Thirty-one (74%) women were in a relationship: 14 had a boyfriend, six lived with their partner and 11 were married. The parity of these 42 women is shown in Table 1. Thirty-three (79%) women were sexually active at the time of the survey.

When asked about their plans for pregnancy, 13 (31%) indicated that they would like to conceive soon (within the next 5 years), five (12%) would like a child at some time in the future (in 5–10 years time) and 15 (36%) wanted to avoid pregnancy. Of these, five never wanted children, five were worried that their health might deteriorate if they became pregnant and five said that their family was complete. A further nine (21%) women were unsure whether they ever wanted to become pregnant.

Thirteen of the 42 responders had had 19 pregnancies between them and of these five (26%) pregnancies were unplanned. Of the five unplanned pregnancies, two continued to term and three were aborted. The case notes of these five women with an unplanned pregnancy were reviewed to determine whether contraception was being used at the time of conception. There was no information

**Table 1** Parity of the study participants

Parity	Patients [n (%)]	
Nulliparous	29	(69)
0+1 (termination)	3	(7)
1+0	5	(12)
2+0	3	(7)
2+1 (miscarriage)	1	(2.5)
2+2 (ectopic/miscarriage)	1	(2.5)

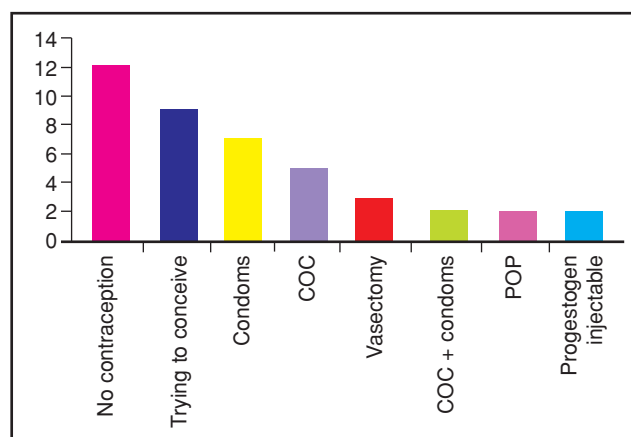
**Table 2** Number of contraceptive methods used by the study participants

Contraceptive methods used (n)	Women [n (%)]	
0	0	(0)
1	13	(31)
2	16	(38)
3	9	(22)
4	3	(7)
5	1	(2)

for two of the women. For the third, a detailed consultation recorded the risks of pregnancy in relation to her ill health and her contraceptive options. Unfortunately she had not renewed her combined oral contraceptive pill (COC) prescription and had conceived in the interim period. The fourth woman was known to be fertile as she had had an abortion in the past and there was documentation of a family planning consultation in her notes. She chose to continue with this unplanned pregnancy. The fifth woman "did not think she could become pregnant having CF". There was no documentation of a contraceptive method failure. Following their unplanned pregnancies, two of the five women's partners underwent vasectomy. One woman was using condoms for contraception, one woman was using the COC and one woman was using no contraception despite being sexually active.

All the women were asked about their current use of contraception. Their responses are shown in Figure 1. The length of contraceptive usage was variable with a mean of 26 months but one woman had been using a COC for 18 years. All women surveyed had tried at least one method of contraception at some stage. The number of methods tried by each patient is shown in Table 2. Overall, 60% of the women had taken the COC at some time, 52% had used condoms, 17% had tried the progestogen injectable, and 14% of the women had used a progestogen-only pill (POP). Twenty-five (60%) women experienced no adverse effects from their contraception but 17 (40%) reported symptoms such as weight gain, irregular bleeding with POPs and with a COC, headache with the contraceptive implant, mood swings with COC and low libido with condoms. No woman reported a deterioration of her CF symptoms with any one contraceptive method and this was confirmed by the CF specialist (S.B.).

Women were asked when they had first received sexual health advice. This ranged between 11 and 27 years of age, with an average of 16 years. They were then asked at what



**Figure 1** Contraceptive methods used by the study participants at the time of the survey. COC, combined oral contraceptive pill; POP, progestogen-only pill.

**Table 3** Specific advice given to the study participants

Whether advice given	Specific advice [n (%)]		Antibiotics advice [n (%)]	
Yes	14	(33)	16	(38)
No	26	(62)	24	(57)
Don't know	2	(5)	2	(5)

age they would have liked to have been given this information. This was earlier, ranging between 11 and 18 years, with an average of 13.7 years. Most of the initial sexual health information was provided by their general practitioner (GP) (26%), but parents were actively involved in 21% of cases and the CF clinic in 17%. Other sources of information were friends (10%), family planning clinics (FPCs) (10%) and school (7%). Sources of contraceptive advice at the time of the survey were different, with 50% consulting their GP, 17% attending FPCs and 9.5% reporting help from the CF clinic. Most women surveyed thought that the quality of the advice given was "OK" (55%) and 24% thought that the advice they received was very good. Six (14%) women said they had not been given enough advice and three (7%) said they had received no advice about contraception and sexual health.

Seven women made additional comments that the information given by their GP and at the CF clinic was limited or inadequate. Twenty-six (62%) women said that they had not received specific advice about the potential effect that contraception may have on CF and its associated complications. Twenty-four (57%) women said that they had not been given information about the possible interactions between use of antibiotics and combined contraceptive methods.

The advice given to the five women who had an unplanned pregnancy is shown in Table 3. They received their advice from their GP (2), the FPC (2) and CF clinic (1). Only one of these five women felt that she had not received enough general contraception advice, however four of the five women said they had had no specific advice about CF and contraception or the effect of antibiotics on combined contraceptive efficacy.

## Discussion

This survey suggests that women with CF have a relatively high rate of unplanned pregnancy and that they receive suboptimal sexual health advice. No one was using a contraceptive implant, intrauterine contraceptive or female sterilisation as their method of birth control and only a minority had attended a specialist contraception and sexual health clinic. It may be that clinicians are reluctant to suggest female sterilisation, as it would involve a general anaesthetic in women with severe respiratory problems. There may also be a reluctance to suggest a permanent method of contraception in young, nulliparous women, despite the severity of their condition. Vasectomy was chosen as the permanent method of contraception by two couples after their unplanned pregnancies.

Early publications suggested a low rate of pregnancy in women with CF for a variety of reasons.<sup>5-8</sup> Emphasis was placed on thickened cervical mucus acting as a potential barrier to sperm.<sup>9</sup> However, despite their medical problems, most women with CF are fertile. Women with CF need to be aware that, in contrast to men with CF, they require effective contraception if they wish to avoid pregnancy. In this study some women were sexually active, not using contraception and not trying to become pregnant, thus putting themselves at risk of an unplanned pregnancy. A study of contraception usage in the general population

suggested that 25% of women of reproductive age are not using any form of contraception, compared to 50% of women in this study.<sup>10</sup> Despite the higher percentage of women at risk, the survey population had an unplanned pregnancy rate of 26.3%, which is similar to the rate in the general population (30%).<sup>10</sup>

There are some limitations to this study, which include the small sample size. CF is an uncommon condition and the survey was confined to one regional CF centre. However the questionnaire response rate was good (76%) and women reported that it was easy to complete. Perhaps some failed to respond because the survey was three pages long and the questions were of a personal nature, however most women had no reservations in answering all questions and all returned questionnaires were complete.

Women with CF are healthier than in the past and therefore specific issues relating to contraception need to be addressed. Women without CF can consider all contraceptive options. However, those with CF have potential medical complications that include an increased risk of venous thromboembolism, concurrent use of antibiotics, osteoporosis, diabetes, liver disease and cholelithiasis. These conditions are relative contraindications to some forms of contraception.<sup>11</sup> The potential risks of using a particular method must be balanced against the risk of pregnancy and the additional stresses that motherhood places on the woman's health.

Many of these women have implanted venous access systems and may have a small risk of thrombosis. Of greater relevance is their frequent use of additional courses of antibiotics, which might reduce the efficacy of the COC.<sup>12</sup> The number of women in the survey using the COC was low at 12%, compared to 21% in the general population. Condom usage was only 17% in the women surveyed compared to 21% in the general population.<sup>10</sup> However, injectable progestogen and POP use was similar to that in the general population. Women with CF are also at risk of low bone mineral density such that injectable progestogens may not be the most appropriate method, even though this method has high efficacy and good compliance.<sup>13-15</sup>

No major adverse events related to contraception were reported by the women in this study. However, 40% had minor side effects such as headaches, irregular bleeding and mood swings. These problems did not lead to discontinuation of the chosen method of contraception.

During a short course of broad-spectrum antibiotics (<3 weeks), women taking the COC are advised to use additional contraception (such as condoms) or to abstain from intercourse during this time and for an additional week following the course.<sup>11</sup> Many women with CF take antibiotics on a long-term basis with the dose and type changing frequently, thus doctors may be reluctant to prescribe a COC as a first-line method of contraception. However, if women are taking the same antibiotic long term the gut flora will adapt and normal re-absorption will be reinstated.<sup>11,12,16</sup>

To reduce concerns about enterohepatic re-absorption of estrogen, a POP is a useful alternative. Newer POPs such as the desogestrel POP have a 12-hour 'safety' window and, in common with other progestogen-only methods, their efficacy is not affected by broad-spectrum antibiotics. The progestogen-only implant and copper-bearing intrauterine devices (IUDs) or the intrauterine system (IUS) are also suitable alternatives and do not adversely affect CF. The CF physicians were cautious about considering intrauterine methods for women with CF due to their perceived worries that these methods were associated with an increased risk of infection.

Most girls receive sexual health and contraception advice at school at the age of 12–13 years and the average age of first intercourse in the general population is 17 years.<sup>10</sup> Over 32% of 14–15-year-old girls have had intercourse.<sup>15,17</sup> In this survey women received sexual health advice in their late teens, but would have preferred to have received advice earlier. However, no unplanned pregnancies occurred before contraceptive advice was given, suggesting that the age of provision of sexual health information is not the most important factor in preventing unwanted pregnancies. Discussions with the specialist CF nurses highlighted the observation that ‘specialists’ and parents often view sick young adolescents as ‘children’ and do not see that they are developing into young adults.

The patient’s GP or CF team were the main sources of advice and information regarding contraception and sexual health. However, the CF team lacked knowledge and training in the full range of modern contraceptive methods. Only one-third of the women surveyed had ever attended specialist contraceptive services. This may explain why none of the study participants currently used an IUD, IUS or Implanon®. Although it is the practice of the CF teams to offer patients an appointment at the hospital-based contraception and sexual health clinic,<sup>17</sup> most patients decline or fail to attend this appointment, and many state that they would rather see their GP. Patients with CF have to attend many clinics and may not wish to attend further appointments at a different time or location. Provision of accurate and detailed written information would help CF staff, contraceptive services and the patients’ GPs to provide up-to-date advice about contraception options, potential drug interactions, fertility and sexual health. Information on how to access contraceptive and sexual health services across the region could also be included.

Unfortunately at present there is no reproductive health guidance available for women with CF and there have been conflicting opinions in the medical literature over the years. A simple, easy-to-understand leaflet to supplement the fpa (Family Planning Association) leaflets would give specific advice for those with CF and help CF specialists to advise women appropriately.

Provision of specialist sexual health clinics at the same time and in the same location as CF services would allow discussion about contraception, fertility and pregnancy options alongside consultations with the CF physician. The most appropriate time for this would be in the early teens, before these young women become sexually active.

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## JOURNAL REVIEW

**Sensitivity and specificity of multimodal and ultrasound screening for ovarian cancer, and stage distribution of detected cancers: results of the prevalence screen of the UK Collaborative Trial of Ovarian Cancer Screening (UKCTOCS).** Menon U, Gentry-Maharaj A, Hallett R, Ryan A, Burnell M, Sharma A, *et al.* *Lancet Oncol* 2009; **10**: 327–340

Epithelial ovarian cancer (EOC) remains a disease with a poor prognosis and little has been achieved in the preceding decade to improve outcomes in women affected by this malignancy. One potential strategy to reduce mortality from EOC is to detect the disease at an earlier stage, given that women with FIGO Stage I disease have >90% 5-year survival whereas 5-year survival is under 30% in patients with Stages III and IV. The UK Collaborative Trial of Ovarian Cancer Screening (UKCTOCS) set out to examine whether screening for ovarian cancer will reduce mortality from ovarian cancer. It is hugely impressive in its scale and will surely

provide an answer. This paper, however, is a report of the performances of the two testing schedules used in the prevalence screen. Over 100 000 postmenopausal women were randomised to screening with either annual ultrasound (USS) or multimodal screening (MMS) using ultrasound and CA125 (measured as an undisclosed algorithm, rather than a single test). A further 101 359 women were allocated to a control group whose results were not examined in this publication. In the MMS group, 90.9% were low risk and returned to annual follow-up, however nearly 1 in 10 had an intermediate or high-risk result leading to further testing. Ultimately there were 97 women who proceeded to surgery and 46 of these had benign or no disease at operation or final histology. In the USS group, 88% were low risk, while 12% required further testing, leading to 845 operations of which 92% were benign. The decision for surgery was taken by the individual consultants following “clinical assessment” and it is not clear whether there were uniform

guidelines for clinicians to follow in listing a participant for surgery. The operations in the MMS group more commonly involved laparotomy and major complications arose in both groups; 2.8% (22/787) in the USS group vs 4.3% (2/47) in the MMS group. In cases of EOC that were detected only 50% were early stage. Overall MMS performs significantly better than USS with respect to specificity (99.8% vs 98.2%) but not sensitivity (89.4% vs 84.9%). There were more false-negatives in the USS group (8) than the MMS group (4). Although the authors suggest that these screening tests are feasible, it has yet to be demonstrated that they are cost effective, and eventually that they reduce mortality from EOC, and it will be 2011 at the earliest before these data are made available.

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