The sexual health of adolescents with cystic fibrosis

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Knowledge of normal teenage sexual behaviour and an ability to advise on contraception has become increasingly relevant for the paediatric cystic fibrosis (CF) team.

Young adult patients with CF are now experiencing the benefits of steady improvements in the treatment of their condition. They enjoy better physical health and can expect to live longer than ever before. New techniques for the treatment of infertility are available, though not always easy to access. These contemporary developments have raised patients' expectations of enjoying normal sexual relationships and having children of their own.

Adolescents are likely to have questions about their sexuality and those with CF may have particular concerns about how their condition will affect their sexual well being. Some young people may not feel able to voice these concerns. Teenagers' questions are likely to include:

- Will CF affect me sexually?
- Will I develop at the same time as my friends?
- Will CF affect my menstrual cycle, fertility and pregnancy?
- Will drugs used for CF treatment affect my fertility, pregnancy or contraception?
- Will pregnancy affect CF?
- Will CF affect my choice of contraception?
- What will I do if I think I have caught a sexual infection?

Many paediatric CF clinics now continue to care for patients up to 18 years of age and several studies have suggested that teenage patients and their parents have unmet information needs about their sexual health. Many would prefer to know about the implications for sexual health before 16 years of age: 56% of young men, 57% of women and 92% of parents.[1,2] In one survey the CF clinic was the main source of sexual advice for 24% of adolescent patients and 42% of parents,[2] but patients themselves were unlikely to initiate the discussion without the active participation of the CF team.

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TEENAGE SEXUAL ACTIVITY

Moderate delay in the onset of puberty and menarche are recognized in CF, with a mean delay of 2 years in young women and 1.5 years in men.[3] Delayed hypothalamic release of gonadotrophin releasing hormone may explain the delay, which is experienced even by healthy well-nourished individuals with CF.[4]

Despite this modest delay in puberty, the onset of sexual activity in the CF teenager and sexual libido is no different from their peers.[5,6] They are therefore exposed to the same risks of sexually transmitted infections (STIs) and unplanned pregnancies as others in their age group. One Australian study highlighted higher levels of unprotected sex by women with CF than matched controls.[5]

Adolescent sexual activity and risk taking are reflected by high rates of STIs and pregnancy among this age group. Teenage pregnancy rates in England and Wales are the highest in Western Europe.[7] In 2000, the conception rate amongst females aged 15–19 was 63/1000 and amongst 13–15 year-olds 9/1000; 39% of conceptions in those under 20 years of age were terminated by abortion.[7] STIs are common and increasing in incidence. The overall number diagnosed in genitourinary clinics in the UK has more than doubled since 1990. Young people bear the burden of acute STIs and rates of infection are increasing disproportionately in this age group. In 2003 diagnosis of Chlamydia, the most commonly diagnosed bacterial STI, were highest in 16–19 year-old females (1341/100 000) and 40% of females diagnosed with gonorrhoea were aged under 20 years. In males, rates of these infections were highest in the 20–24 age group.[8,9]

CF teenagers would be expected to respond normally to treatment for the commonly transmitted sexual infections such as chlamydia and gonorrhoea though there are no specific data on this subject. Anecdotal reports of chronic lung infection in HIV seropositive patients show a poor outcome with Aspergillus, Pseudomonas, and tuberculous infections predominating.[10,11]

It is recognized that if sex is unplanned it is likely to be unprotected. Among 16–24 year olds, 37% use contraception if sex occurred on the spur of the moment compared with 70% who use contraception when sex is planned ahead. Those who have their first sexual intercourse under the age of 16 were least likely to have used contraception. Significant factors affecting this were

anxiety about confidentiality of the accessed health services, and the unplanned nature of the event.[7]

INFERTILITY IN MALES WITH CF

Adolescent males with CF need to know that, with rare exceptions, they are azo-ospermic due to absence of the vas deferens regardless of the severity of their respiratory or gastrointestinal disease.[12,13] Additionally, there is often absence or dysfunction of the seminal vesicles accounting for the low volume ejaculates.[14] Sexual potency is not affected by these abnormalities but the patient will need reassurance on this issue as a number of misunderstandings have been reported.[6]

The importance of infertility assumes greater magnitude with increasing maturity and this issue may require revisiting on more than one occasion. Semen analysis is not routinely available in CF clinics for the 2% of males who may be fertile, and should not be undertaken within a few months of serious illness. There are no normal data for sperm counts in males under 18.

Current reproductive options for men with CF are artificial insemination of their partner by donor sperm, or surgical retrieval of their own sperm using the techniques of percutaneous epididymal sperm aspiration (PESA) or testicular sperm extraction (TESA). Surgically retrieved sperm are immature and incapable of fertilization by conventional means, so fertilization is achieved by injection of a selected sperm into the cytoplasm of a mature oocyte (intracytoplasmic sperm injection, ICSI).[15] This technique achieves significant fertilization rates with live birth rates of 15–55% reported.[16,17] More than 26 000 babies have been born following the introduction of ICSI in 1992 with an overall live birth rate of nearly 30% of transferred embryos.[17] Reports of major birth defects occurring twice as frequently as in the background population,[18,19] together with concerns that genetic disorders in the father may be easily passed on to the offspring suggest that counselling and genetic screening should be mandatory.[20]

Partners should be screened for the cystic fibrosis transmembrane conductance regulator (CFTR) mutation because of the increased risk of CF in the offspring, with pre-implantation embryo diagnosis if the female partner carries a mutation. There is a 1 in 50 risk of an affected child with an unscreened partner.

The lengthy waiting list for these newer infertility treatments may mean that men miss out on several valuable years of fatherhood unless they opt for expensive privately funded treatments. Some have reported anecdotally that they would have given a high priority to funding infertility treatments if they had been informed about this treatment option in adolescence.

In the UK there have been no successful applications for adoption made by CF males because of their reduced life expectancy.

INFERTILITY IN CF WOMEN

Earlier reports of relative infertility in women with CF[21] are now misleading. Those with good growth and lung function can expect to develop into sexually mature adults with normal ovulation.

Although cervical mucous is unusually tenacious and does not show changes in hydration during the menstrual cycle,[22] there is evidence of near normal fertility with increasing numbers of pregnancies reported each year. Pregnancy rates in CF are probably under-estimated but available data suggest a prevalence of 40 pregnancies per 1000 menstruating women over 16 years of age compared with 80/1000 in healthy women.[23] Contraceptive precautions are advisable for all sexually active young women with CF until they wish to plan for pregnancy.

CONTRACEPTION

Sexual encounters between teenagers are frequently unplanned and the partner may be unfamiliar so the ideal contraceptive should achieve the dual aims of avoiding unplanned pregnancy and prevention of STI. Although condoms are widely available the reported user failure rate of 10% by young users may be unacceptable and additional contraceptive precautions advisable.

The extensive range of contraceptive methods in current use include abstinence, withdrawal, barriers (diaphragms, condoms, femidoms), hormonal pills (combined and progestogen-only), intrauterine devices and systems (IUD and IUS), injectables, implants, patches and sterilization. Some are 'natural' methods, some are available 'over the counter' and others require access to medical services. Discussion of all the methods of contraception, their contraindications and side effects is out-with the scope of this paper. Clinicians are referred to the faculty of family planning and reproductive healthcare website www.Faculty of Family Planning and Reproductive Health Care.org.uk and the World Health Organization (WHO) *Medical Eligibility Criteria for Contraceptive Use* which provides evidence based guidance.[24]

The young woman with CF who requires safe and effective contraception is advised to consult a clinician experienced in family planning and reproductive healthcare with whom she can discuss the risks and benefits of available methods. Collaboration between the family planning clinician and the teenager's CF physician is recommended.

What methods of contraception are suitable for use by women with CF?

Neither the Faculty of Family Planning and Reproductive Healthcare, nor the Royal College of Obstetricians and Gynaecologists (RCOG) currently provide guidance for contraception for women with CF. A search of the medical literature from MEDLINE, PubMed, EMBASE and Cochrane databases found very few papers relating to contraceptive use in women with CF and those that were found express conflicting opinions.[25–28]

An enquiry made to the clinical effectiveness unit of the faculty of family planning and reproductive healthcare met with the following response:

> 'There is no specific reference to cystic fibrosis in the World Health Organization *Medical Eligibility Criteria for Contraceptive Use*. However, patients with cystic fibrosis may suffer from gall bladder disease due to viscous secretions. World Health Organization medical eligibility criteria for contraceptive use recommends that for asymptomatic women with gall bladder disease, the benefits of the combined oral contraceptive pill, progestogen-only, depot medroxyprogesterone acetate, and the levonorgestrel-releasing intrauterine system outweigh the risks (WHO category 2). For symptomatic women who have current or medically treated gall-bladder disease, the risks of the combined oral contraceptive pill outweigh the benefits (WHO category 3). Following a cholecystectomy women are classified as benefits outweigh risks. For symptomatic women with current or medically treated gall bladder disease, the benefits of the progestogen-only pill, depot medroxy-progesterone acetate and levonorgestrel-releasing intrauterine system outweigh the risks. Both symptomatic and asymptomatic women with gall-bladder disease may have unrestricted use of the intrauterine device. The Clinical Effectiveness Unit found no evidence on the use of the combined contraceptive patch by women with CF' (Faculty of Family Planning and Reproductive Healthcare Clinical Effectiveness Unit Members Enquiry Service, reference 716).

It seems from the literature that there is no particular method of contraception contra-indicated or recommended for women with CF. Each method needs to be weighed up against the particular pathologies, caused by cystic fibrosis, that are present in the woman, as one would do with all patients.

Contraceptive options for women with CF who fulfil the WHO medical eligibility criteria for contraceptive use are the same as for those without this condition. Whilst there is no specific recommendation for contraceptive use in adolescents with CF, important considerations may include risk of venous thromboembolism, drug interactions, diabetes, active liver disease, severe pulmonary hypertension, osteoporosis and cholelithiasis. Risks and benefits of all contraceptive methods should be assessed on an individual basis. Any risk associated with a contraceptive method must be weighed up against the risks of unplanned pregnancy, which for women with CF can be significant (Tables 1 and 2).

Combined oral contraceptive pill

The combined oral contraceptive pill is widely used by sexually active teenagers and young women in the UK. It inhibits ovulation and acts on cervical mucus to prevent pregnancy. It is highly effective, with method failure rates of 0.1 per 100 woman years, although typical user failure rates are quoted as 5 per 100 woman-years.[29] Its ease of use, reversibility, and menstrual control make it a popular method. Non-contraceptive benefits include regulation of menstrual bleeding and, for some, improvements to acne. However, it is dependent on user compliance and is unsuitable for those for whom oestrogen is contra-indicated. It can be used by women who fulfil the WHO medical eligibility criteria for contraceptive use.[24] Absorption and efficacy of combined oral contraceptive pill in women with CF appears to be as good as in healthy women.[29] Only one small study investigating the impact of combined oral contraceptive pills on pulmonary function in adolescent and young women with CF and moderate-to-severe obstructive lung disease was identified; no significant deterioration in clinical status or pulmonary function was found.[30] Some authorities recommend the combined oral contraceptive pill as the method of choice to young women with CF.[31] The main concerns about its use by women with CF relate to venous thromboembolism, cholelithiasis (see above) and to co-administration of antibiotics.

Venous thromboembolism

Women should be advised that the risk of venous thromboembolism increases with combined oral contraceptive pill use. The risk to users of a second-generation combined oral contraceptive pill (containing levonorgestrel or norethisterone) is increased threefold over the non-user rate to 15/100 000. Use of third generation combined oral contraceptive pill (containing desogestrel or gestodene) increases the absolute risk to 25/100 000.[24] Any increase in risk must be compared to the risk of venous thromboembolism associated with pregnancy which is 60/100 000. The combined oral contraceptive pill is contraindicated for women with a personal history of venous thromboembolism, known thrombogenic mutations (e.g. Factor V Leiden; Prothrombin mutation; Protein S, Protein C, and Antithrombin deficiencies) or undergoing major surgery

Table 1 **Pathologies and treatments associated with cystic fibrosis that influence contraceptive choice** [Adapted from WHO *Medical Eligibility for Contraceptive Use*, 3rd edn (Ref. 24)]

	COC	***Evra***	***POP***	***DMPA***	***Implanon***
Deficits in bone mineral density	No negative effect. May have positive effect		No evidence of effect	Negative effect on bone mineral density. Should not be used	No evidence of effect
Diabetes Non-vascular disease	Can be used: benefits outweigh risks				
Nephropathy/ retinopathy/ neuropathy	Should not be used: unacceptable health risk		Can be used: benefits outweigh risks	Risks usually outweigh advantages: can be used with caution	Can be used: benefits outweigh risks
Cholelithiasis asymptomatic and post-cholecystectomy	Can be used: benefits outweigh risks				
Cholelithiasis symptomatic Medically treated Current	Can be used with caution but risks usually outweigh advantages		Can be used: benefits outweigh risks		
Active liver disease	Absolute contraindication		Can be used unless severe changes		
Pulmonary hypertension	Absolute contraindication		Can be used		
Implanted venous access device	Relative contraindication. Assess risk		Can be used		
Interaction with broad spectrum non-enzyme inducing antibiotics	Yes: see text. But does not restrict use	No data	No interaction		
Interaction with liver enzyme inducing drugs	Significant effect: see text	No data: advise as per combined oral contraceptive pill	Significant effect: see text	Theoretical risk	Significant effect: see text

COC, combined oral contraceptive pill; Evra®, combined contraceptive patch; POP, progestogen-only pill; DMPA, depot medroxyprogestogen acetate

with immobilization.[32] There is no evidence of a hypercoagulable state inherent to CF. Catheter related venous thrombosis is however a relevant complication reported in 5–14% of patients with totally implantable vascular access devices, widely used for intermittent administration of intravenous antibiotics.[33–35] The risks of thrombosis must be carefully assessed if a woman has a totally implanted venous access device in situ and wishes to take the combined oral contraceptive pill; safer alternatives should be considered.

Drug interactions

Drugs known to have a clinically significant impact on contraceptive efficacy include rifampicin and rifamycin, griseofulvin, some anticonvulsants (topiramate, barbiturates, carbamazepine, and primidone) and in some women short courses of tetracyclines and ampicillin.[36] The interaction with broad spectrum antibiotics is due to an effect on the entero-hepatic circulation of the ethinyloestradiol component of the combined pill. Short-term (<3 weeks) broad-spectrum antibiotics may alter gut flora and pregnancies have been reported following their use in women using combined oral contraceptive pills.[32] Gut flora are reconstituted with resistant organisms in about 2 weeks, so it is in the use of short-term antibiotic therapy, or in long-term therapy at the time of change to a new antibiotic, that concern about efficacy of combined oral contraceptive pill arises.[37] Women using combined oral contraceptive pill should use additional contraception while taking non-enzyme inducing broad-spectrum antibiotic courses of less than 3 weeks and for 7 days after stopping; the pill-free interval should be omitted if the first 2 weeks' antibiotic use extended into the last 7 days of a pack ('7 day rule'). Women who are established on a non-enzyme-inducing antibiotic long-term (more than 3 weeks) do not require additional contraception unless they change to a different broad-spectrum antibiotic.[32] When a broad

Table 2 **Advantages and disadvantages of some contraceptives used by teenagers**

Contraceptive	*Efficacy*	*Advantages*	*Main disadvantages*
Combined oral contraceptive pill	>99% when used correctly	May reduce bleeding, period pain and pre-menstrual tension. May improve acne vulgaris. Contraceptive effect can be terminated by woman herself at any time. Reduces risk of ovarian and endometrial cancer	Requires prescription. Increased risk of venous thromboembolism. Interaction with broad spectrum antibiotics, and liver enzyme inducing drugs. Not effective if taken over 12 h late. Not suitable for those for whom oestrogen is contraindicated.
Progestogen-only pills	96–99% when used correctly	Fewer side effects than combined oral contraceptive pill. No need for extra precautions with non-enzyme inducing antibiotics. Contraceptive effect can be terminated by woman herself at any time	Requires prescription. Most demand punctuality in pill taking. Irregular bleeding patterns. Interaction with liver enzyme inducing drugs
Progestogen only implants (Implanon®)	>99%	Effective and rapidly reversible. Lasts up to 3 years. Immediately reversible. Independent of user compliance	Insertion and removal requires trained professionals. Irregular bleeding patterns. Interaction with liver enzyme inducing drugs
Progestogen injection (Depo-Provera®)	>99%	Useful for women who forget to take tablets. Administered at 12 week intervals	Requires administration by trained practitioner. Weight changes in some women. Significant effect on bone mineral density. Bleeding disturbance initially in some users. Cannot be immediately reversed. Can take time for fertility to return. Some complain of acne
Intrauterine device	98–99%	Lasts up to 10 years (depending on type). Effectiveness is independent of user. Nulliparity is not a contraindication	Insertion and removal requires trained professionals. Some experience heavy, painful, prolonged menses
Combined contraceptive patch	>99% when used correctly	Once-weekly. Contraceptive effect can be terminated by woman herself at any time	Requires prescription. Demands user compliance. Not suitable for those for whom oestrogen is contraindicated. Interaction with liver enzyme inducing drugs
Condoms	92–96% but less if include user error	Widely available. No need for medical involvement. Non-hormonal protection against sexually transmitted infection	Must be acceptable to both partners. May come off or split. Efficacy dependent on careful use

spectrum antibiotic which is intended for long-term use is first introduced to a woman established on the combined oral contraceptive pill, extra precautions need to be sustained for the first 2 weeks, plus the usual 7 day rule.[37]

Other drug interactions with combined oral contraceptive pill

Drug-induced stimulation of hepatic cyp450 enzymes lead to increased metabolism and elimination of both oestrogen and progestogen so may significantly affect efficacy of hormonal methods of contraception. The interaction with rifampicin is clinically significant. Women using oral hormonal contraception (combined or progestogen only pills) and who need to take a short course of rifampicin (1 week or less) should be advised that they will need to take extra contraceptive precautions as soon as they start taking rifampicin and for 4 weeks after the last dose; they should take their pill continuously, omitting any pill-free interval whilst taking rifampicin and during the 7 days after the last dose. Those who need to take a long course of rifampicin should be advised to choose an alternative contraceptive method; the risk of hormonal contraceptive failure and consequent unplanned pregnancy is high.[38] If a woman using a liver enzyme-inducing drug still chooses to use combined oral contraceptive pill, established UK practice is to use a regimen containing at least 50 μg of ethinyloestradiol daily and a barrier method of contraception. Additional contraception is also required for 28 days after the enzyme-inducer is stopped.[32]

Some complementary medicines, notably St John's wort, can potentially reduce efficacy of oral contraception due to the effect on liver enzymes. Some drugs may have their bioavailability increased by interaction with combined oral contraceptive pill with potentially toxic effects; these include theophylline and cyclosporin.[37]

Pulmonary hypertension

The combined oral contraceptive pill is absolutely contraindicated in patients with pulmonary hypertension.

Progestogen only methods

Women with CF who fulfil medical eligibility criteria can use progestogen-only methods. They may appeal especially to those for whom exogenous oestrogen is contraindicated and may be used by women with a personal history of venous thromboembolism or immobility. These methods are suitable for patients with pulmonary hypertension. Progestogen does not undergo an entero-hepatic circulation so additional contraception is not required when women who are using progestogen-only methods are prescribed non-enzyme-inducing antibiotics for any duration. However, with the possible exception of Depo-Provera, progestogen-only methods are affected by liver enzyme inducing drugs.

Progestogen-only pill

Progestogen-only pills act by altering cervical mucus and, in some women, inhibiting ovulation. The overall failure rate is 0.3–4/100 woman years. With the exception of Cerazette® (see below) the progestogen-only pill must be taken with extreme regularity.[37] Compliance in the young may be problematic and the expected failure rate in teenagers is likely to be in the order of 4%. Its efficacy in young women with CF is unknown. Concern that progesterone could cause bronchial mucus thickening and lead to impaired lung function[25] has not been substantiated. Studies have not found any evidence for deterioration in lung function or an increase in acute pulmonary exacerbations in CF patients taking exogenous progestogen.[31]

An interesting recent addition to this group of pills is Cerazette® (Organon Laboratories Ltd), a desogestrel-only pill. Its primary mode of action is inhibition of ovulation and it also causes changes in cervical mucus. Its manufacturers promote it as the first oestrogen-free pill with the efficacy of the combined oral contraceptive pill. It is not recommended as an alternative to combined oral contraceptive pill in routine practice, but provides a useful alternative for women who require oestrogen-free contraception.[39] Cerazette might prove useful for women with CF as non-liver-enzyme-inducing antibiotics do not affect its efficacy and women with risks of venous thromboembolism can use it with caution. Organon advises its discontinuation in the event of thrombosis. It is more forgiving than other progestogen-only pills to those who are imprecise in timing of pill taking, which may benefit younger women. Its effects on bone mineral density are unknown though oestradiol levels are maintained above levels found in the early follicular phase of ovulation[40] suggesting that bone mineralization is protected. Bone density was not affected over a 2-year use of Implanon, which releases the biologically active form of desogestrel.[41]

Injectable methods

Depot medroxyprogesterone acetate (Depo-Provera®) is a highly effective method of contraception with a failure rate $<1\%$. It acts by suppressing ovarian activity and inhibiting ovulation. No studies were identified which investigated the use of depot medroxyprogesterone acetate by women with CF. However, clinicians may be cautious about its use in these women due to concern about osteoporosis. Reports of an association between long-term depot medroxyprogesterone acetate and osteoporosis in healthy women are conflicting. In healthy teenage users current evidence indicates a reduction in bone mineral density with depot medroxyprogesterone acetate use at a time when bone mineral density is normally increasing. In this age group the Committee on Safety of Medicines now recommends alternative contraceptive methods where there are significant risk factors for osteoporosis.[42] Risk of osteoporosis is increased in those with personal risk factors including amenorrhoea, low body mass index, corticosteroid use, thyroid disease and family history.[43] As deficits in bone mineral density resulting in premature osteopenia and osteoporosis have been well documented in patients with cystic fibrosis[44] caution must be advised if a young woman with CF wishes to use depot medroxyprogesterone acetate. Treatment should only be considered after other methods have proved unsuitable, and risks of treatment must be reviewed within 2 years. Dual energy X-ray absorptiometry (DEXA) would seem prudent if a young woman with CF wishes to use depot medroxyprogesterone acetate.

There is no correlation between amenorrhoea and hypoestrogenism. Oestradiol levels are not generally recommended and should only be considered if the woman has been using depot medroxyprogesterone acetate for 5 or more years with or without amenorrhoea or at any time if she has symptoms suggestive of hypo-oestrogenism.

Implanon® or Cerazette are alternative progestogen-only options where there is concern about osteoporosis, as although they inhibit ovulation, ovarian activity is not completely suppressed so that oestrogen deficiency is unlikely despite amenorrhoea (see below).

Progestogen-only implants

Sub-dermal contraceptive implants provide long-acting, highly-effective and reversible contraception by achieving low and stable concentrations of synthetic progestogens. No pregnancies were reported during clinical trials. Young women with CF who fulfil medical eligibility criteria can use the progestogen-only implant (Implanon). Women who have contraindications to oestrogen can use this method. It is unaffected by antibiotics but like other hormonal methods its efficacy is significantly affected by liver enzyme inducing drugs. The manufacturers advise removal in the event of a

thrombosis and prior to long-term immobilization due to surgery or illness. It can be used with caution in women with a history of, or risk factors for, venous thromboembolism. Fecundity returns immediately following removal. Its insertion and removal are minor surgical procedures, which carry the risk of infection and bleeding. Careful counselling is needed prior to insertion about potential side effects, particularly irregular bleeding patterns.[45]

Copper-containing intrauterine device

Young women who fulfil the medical eligibility criteria can use an IUD, although its use in this age group is not common. Its benefits to the sexually active teenager with CF are its low failure rate, suitability for those with a personal history of venous thromboembolism or immobility, lack of interaction with drugs and it does not affect osteoporosis. Its efficacy does not depend on ongoing action by the young woman and it is reversible. It can be used by nulliparous women. The disadvantages include an increased risk of pelvic inflammatory disease in the first 21 days following insertion[46] and bleeding problems, which are troublesome for some women.

Barrier methods

Barrier methods include condoms, femidoms and diaphragms. User failure rate may make these unacceptable for young people. Everyone who is sexually active should be aware that the pill, coil, injection, implant, etc., can protect against pregnancy but not STIs, and should be encouraged to use condoms appropriately. Women of all ages should be given information about emergency contraception and how to access it.

Emergency contraception

Emergency contraception[47] provides a safe means of preventing pregnancy following unprotected sexual intercourse or potential contraceptive failure. Two methods are currently available. The first, which is more widely accessible, is the progestogen-only emergency contraception method which comprises a single dose of levonorgestrel 1.5 g available as Levonelle-2®/Levonelle®. Progestogen-only emergency contraception can be obtained free on prescription, free under Patient Group Directions from some pharmacists and over the counter at a cost (currently £25) from pharmacists (who cannot sell it to under 16s). The regimen for progestogen-only emergency contraception use by a woman with CF is the same as in healthy women. Progestogen-only emergency contraception efficacy may be significantly affected by liver enzyme inducing drugs. The other method of emergency contraception is a copper intrauterine device which is almost 100% effective when used appropriately.

PLANNING FOR PREGNANCY

Young women with relatively mild lung disease (FEV1 $>70\%$ predicted) can be reassured that pregnancy should be well tolerated with reasonable expectations of delivering a healthy baby.[48] Women with more severe lung disease may show a worrying decline in respiratory status as pregnancy progresses with increased risks of premature delivery or termination to protect the mother's health.[49] Approximately 80% of pregnancies will result in delivery of a live baby.

Pregnancy will have the best outcome when it is planned with close collaboration between obstetric and CF teams. Pre-pregnancy advice should include genetic counselling and carrier screening of partners because of the 1 in 50 risk of an affected child if the mother's status is unknown.

Emphasis must be given to achieving the best possible respiratory function and optimal control of diabetes before and during pregnancy. Different approaches to physiotherapy and exercise regimes may be required. Nutritional supplements are recommended to achieve a weight gain of 12.5 kg during pregnancy. Folic acid supplements should be given before conception and vitamin A supplements should be reduced to not more than the daily recommended intake.[50] A check of the serum retinol level is advisable. The CF team should review maternal medication before and during pregnancy to avoid potentially teratogenic drug effects from aminoglycosides, ursodeoxycholic acid and proton pump inhibitors. Caution is advised in the use of ciprofloxacin, co-trimoxazole, rifampicin, chloramphenicol, and fluconazole, all of which should be avoided if possible.

In optimal circumstances, prospective case control studies have shown that the long-term decline in lung function is not worse in CF women after pregnancy compared with controls and 3 year survival rates are comparable with nulliparous CF women.[51] Poorer outcomes in pregnant cases and controls were associated with poor pre-pregnant lung function, low body weight and diabetes.[52]

Safe management of the birth often involves a planned pre-term delivery.[53] Women with the highest levels of lung function can expect to deliver a live birth at term. Breast feeding is not contra-indicated providing the mother's nutritional status is satisfactory and salt levels in breast milk are normal.[54] Foetal abnormalities are reported but probably occur no more frequently than in the normal population.[55]

An important priority for the CF team is to try to ensure that women with CF are aware of the risks of unplanned pregnancy. However, an important consideration for the

mother will be the better outcome for childbirth at a relatively young age before her lung function has declined to hazardous levels.

MANAGEMENT OF SEXUAL HEALTH BY THE CF TEAM

A holistic approach to contraceptive provision considers an individual's overall sexual and reproductive health needs. It informs young men and women of potential benefits and risks in an understandable way enabling them to take their own informed contraceptive choices. Liaison between healthcare professionals is recommended.

Members of the CF team may have the opportunity to discuss sexual health concerns with young people in their care, to promote sexual well being, address high-risk sexual behaviour and facilitate access to confidential sexual healthcare services. Teenagers who have been exposed to risks of STI should be encouraged to attend a genitourinary medicine clinic for assessment. Support, training and research can help to address the uncertainty among nurses treating adolescents as suggested in the document (2003, Royal College of Paediatrics and Child Health).[56,57] Concern that sex education encourages sexual experimentation is unsubstantiated.

For some adolescents fears about confidentiality and embarrassment may make them reluctant to disclose sexual activity to the clinician who has looked after them throughout childhood. Others may feel that this is the most appropriate person with whom to discuss their sexual health concerns. Disclosure may be discouraged by the presence of parents during a consultation. It is important to be aware that a young woman may have accessed hormonal contraception without the knowledge of either her paediatrician or her general practitioner and may not spontaneously forward this information. This may have significant implications in terms of possible interactions with changing medications. Clinicians will need to ask sensitively about contraceptive use. Young women should be strongly advised to inform their paediatrician about prescribed contraceptive use with appropriate reassurance about confidentiality.

The CF team should provide supportive literature and direct teenagers and their parents towards relevant websites and support groups as necessary. Some CF organizations have produced information booklets about sexual health that can be accessed over the Internet.[58,59]

CONCLUSIONS

Young men and women with CF are not different from their peer group in their sexual behaviour. They are becoming sexually active at an increasingly early age, have higher rates of STI than the general population, and the teenage pregnancy rate in the UK is the highest in Western Europe.

Knowledge about reproductive health among adolescents with CF is poor. The CF team can make a significant contribution to this unmet need through sensitive inquiry, reassurances about confidentiality, and informed advice. Liaison with family planning clinicians and genitourinary clinics is to be encouraged.

Nearly all adolescents with CF can now have realistic expectations of parenthood. They may elect to conceive at a relatively young age when the outcome for their own health is best and when they can maximize their years of caring for their offspring. However, conception is not without special risks and high priority should be given to the provision of contraceptive advice and avoiding unplanned pregnancies.

All contraceptive choices are available for women with CF and no particular method is recommended or contraindicated. In weighing the risks and benefits of prescribed contraceptives the combined oral contraceptive pills are acceptable and widely used; but newer desogestrel containing progestogen-only pills appear to be as effective and associated with fewer potential hazards. Each method needs to be weighed against the woman's existing CF pathologies and with conformity to WHO recommendations. The CF clinician can make an important contribution to achieving the best choice for each individual.

KEY FACTS

- Knowledge about reproductive health amongst adolescents with CF is poor
- The majority of men with CF are infertile due to congenital bilateral absence of the vas deferens
- Female sub-fertility is less common and conception can still occur despite severe disease.

Teenage sex

- More than 1:5 UK teenagers have sex before their 16th birthday
- The UK has the highest teenage pregnancy rate in Western Europe
- STIs are common and increasing in incidence
- Young women bear the brunt of morbidity from STIs
- Asymptomatic infection is common.

Contraception

- Contraceptive options for women with CF who fulfil the WHO *Medical Eligibility Criteria for Contraceptive Use* are the same as for those without this condition
- There is no particular method of contraception contraindicated or recommended for women with CF. Each method needs to be weighed up against the particular

Box 1 **Competency to consent to treatment**

The Fraser criteria (1986) must be met to allow healthcare professionals to provide contraception to people under 16 years of age without parental consent:

- He/she understands the potential benefits/risks of the treatment and advice given
- He/she is likely to have sex without contraception
- His/her physical or mental health is likely to suffer if contraception/advice is not given
- It is in his/her best interest to receive contraception/advice without parental consent
- The practitioner is legally obliged to discuss value of parental support

pathologies, caused by cystic fibrosis, that are present in the woman, as one would do with all patients

- Important considerations may include venous thromboembolism, drug interactions, osteoporosis, diabetes, and cholelithiasis
- Risks and benefits of all contraceptive methods should be assessed on an individual basis and balanced against the risks of unplanned pregnancy.

Access to services

- Barriers to teenagers accessing services include inaccessibility and fears about confidentiality.

Law

- The duty of confidentiality owed to person under the age of 16 is as great as that owed to any other person
- The Fraser criteria[60] must be met to allow healthcare professionals to provide contraception to people under 16 years of age without parental consent (Box 1).

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