

Review

Communication of information about reproductive and sexual health in cystic fibrosis. Patients, parents and caregivers' experience

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Abstract

Background: This review evaluated research concerning communication and information about reproductive and sexual health (RSH) in cystic fibrosis (CF).

Methods: Papers in the English language reporting RSH issues in CF, published between January 2000 and December 2010, were included. The review focused on (a) the content of information given to parents and patients, (b) the timing of information, (c) the sources of information, (d) attitudes and emotional reactions, (e) chronic illness and sexual behavior, (f) methodological and cultural considerations and (g) ethical considerations.

Results: Eleven papers were identified originating from Australia, the United Kingdom and Poland. Patients and parents expressed the need for up-to-date, CF-related verbal and written information, provided by the CF team and infertility specialists. Patients were often embarrassed to initiate a discussion. Health care providers expressed the need for training to counsel patients in RSH.

Conclusion: A pro-active discussion of RSH issues is proposed as a standard part of the care-pathway. A list of recommendations is given to initiate this process.

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Keywords: Cystic fibrosis; Reproductive health; Sexual health and sexuality

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1. Introduction

In 1982, Levine and Stern [1] conducted a study to investigate the sexual functioning of married patients with cystic fibrosis (CF). Nine of the 30 patients (population of 450) reported serious sexual problems and five patients attributed these to their CF. This study was one of the first, concluding that adult patients with CF contemplating marriage have a reasonable chance for normal sexual functioning. Nearly 30 years later, patients still have their expectations of a normal sexual relationship and, above all, hope for children [2,3]. These raised expectations are based on an improved life expectancy and better quality of life and possibly on the improved information patients and/or parents receive about this topic. Since 1982 a number of papers have studied reproductive health in CF [4]. Despite this, little is known about the progress that has been made in informing and educating patients, nor about the different topics that are included within the theme of reproductive and sexual health (RSH) in CF.

The present review will focus on (a) the content of information given to parents and patients, (b) the timing of information, (c) the sources of information for parents and patients, (d) attitudes and emotional reactions in communicating about RSH, (e) chronic illness and sexual behavior, (f) methodological and cultural considerations and (g) ethical considerations. We will conclude with recommendations drawn from the literature and discuss topics that need further study.

2. The identification of papers

PubMed was searched to locate all full papers in the English language reporting reproductive and sexual health issues in CF, published between January 2000 and December 2010. Secondary searches were done using the papers that were identified. Papers were located that included ‘cystic fibrosis,’ ‘reproductive health,’ ‘sexual health’ and ‘sexuality’ in the title, abstract or keywords. A total of 11 research papers were identified, originating from Australia, the United Kingdom and Poland. Table 1 provides a concise overview of these studies.

The study populations were male patients (five papers) [5,8,10,14,15], female patients (one paper) [12], mixed sample of males and females patients (one paper) [6], health care professionals (one paper) [7], mixed sample of parents and female patients (two papers) [9,13] and parents of male patients (one paper) [11].

3. Content of information

The content of information on RSH that patients and parents receive is diverse and seems to depend on patient gender, CF center and/or country. Not all studies clearly distinguish between general and CF-related sexual health information. A list of important topics, based on the recent (medical) literature, is given in Table 2.

Two studies (Polish and UK samples) have concluded that women in both cultures reported suboptimal sexual health advice in relation to CF and its treatments [12,13]. In particular, they had a poor understanding of contraceptive use in relation to antibiotic treatment. According to the UK authors, this resulted

in 50% of women not using any contraception, compared to 25% of healthy women, and resulted in more unplanned pregnancies in the CF group.

Most men with CF seemed aware of both the extent and cause of their infertility [5,6,8,10,15], although they expressed the need for information on reproductive options. Under-informing patients may lead to misconceptions and risky sexual behavior [7,12]. Sawyer et al. [10,15] reported two important misconceptions. One in three men assumed they did not need to use a condom and those informed by health professionals were more likely to assume this than men informed by their parents. One in 10 men mixed up infertility with impotence. Another misinterpretation is reported by Popli et al. [14] with patients overestimating the risk of their biological child having CF after conception with assisted reproductive techniques. The majority of health care professionals routinely informed adolescent boys that sexual performance is not affected by CF, but only 3% inform them about small volume ejaculations [7].

An Australian study [11] noted that parents were well informed, although only 30% of them were satisfied with their current RSH knowledge. All parents knew about the risk of sexually transmitted diseases in both CF and their healthy peers and most understood that their son could father a child. Only 43% of parents were certain their sons (aged 6–19 years) had been informed of probable infertility. The Polish study [13] reported several misunderstandings. Only 23% of mothers of girls understood the problems connected with female fertility and 58% thought that women with CF had absolutely normal fertility. In addition, 44% of this sample believed that men with CF had normal fertility.

4. Timing of informing

Girls more than boys initiated the discussion about fertility issues [6]. Both genders received their first information between the ages of 12 and 16 [9–13,15]. For Australian and Polish girls this was around 13 years of age [9,13]. In the United Kingdom, girls were given contraceptive advice around the age of 16 years although they would have preferred to have been informed around the age of 13 [12]. Australian mothers informed their daughters about RSH issues around 9 years of age [9]. Importantly, girls requested more information with increasing age, probably due to them becoming more interested and/or active in sexual behavior [9].

Sawyer et al. [10] found that boys were rarely informed by health care professionals about infertility before the age of 15. Yet, when parents informed their sons they preferred to do so at an earlier age than the CF doctor (13.2 years versus 17.2 years) [10,15]. An earlier study from the United Kingdom [8] reported that 56% of boys never received advice on infertility or contraception. These results are unexpected as in other studies the majority of boys had been informed about their infertility. The majority of men in all cultures said that it was pertinent to inform boys early, and earlier than is currently being done, preferably during their early teens [5,7,14,15]. Most health care professionals consider 13–14 years the best age to inform male patients about infertility [11]. Studies do not report on professional’s views on the best time to inform female patients about RSH and CF.

Table 1
Studies conducted on reproductive and sexual health (RSH) in cystic fibrosis: Information, attitudes and experiences.

| Authors | Topic | Sample | Method | Results | Main conclusions |
|------------------------|---------------------------------------------------------------------------------------------------------------------------|------------------------------------------------|----------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Rodgers et al. [5] UK | Male infertility: best time to inform | 18 male patients | Questionnaire | Sources of information were the CF team [6], parents [5], and written information [2]. Five discovered the information unexpectedly at a median age of 17. 14/18 expected the CF team to discuss infertility. All patients wanted the CF team to initiate the discussion, supported with written information and repeated over time. 5/18 had semen analysis, 17/18 think this should be routine. | CF patients prefer discussion about infertility at an earlier age than given. Semen analysis should be routine. Need for liaison between pediatric and adult services. |
| Fair et al. [6] UK | Attitudes to fertility and information of men and women with CF | 82 male and 54 female patients | Postal questionnaire | All but two men knew about infertility. For 85% of men and 72% of women having children was important. 43% of men and 26% of women had never had any discussion of fertility issues with CF team. 56% of men think this should occur before the age of 16. Learning about infertility was associated with strong negative emotions. Women were more likely to initiate the discussion. | Need for improvement in information process about infertility. Timing is important. Important gender differences. Health care team should initiate discussion. |
| Sawyer et al. [7] USA | Reproductive and sexual health in males with CF, attitudes of health care professionals: Practice, attitudes and barriers | 32 health care professionals from 4 CF centers | Interview | 66% informed parents of male patients at diagnosis 22% informed during late childhood. All discussed with adolescents. Age thought appropriate was 13.8, but actual information was around age 15.2. 50% inform that sexual performance is not affected by CF. 38% inform about condoms. 50% discuss normal sexual performance. 13% offer semen analysis. 3% inform about small volume ejaculation. 19% discuss reproductive options. Barriers reported were embarrassment, time, insufficient training. | Health care professionals are aware of need for information. Embarrassment and time were major barriers. Need for training. |
| Thickett et al. [8] UK | Awareness of infertility in men with CF | 72 male patients | Questionnaire | 56% never received advice on infertility or contraception. 24% received information from CF clinician, Remainder received information from parents, other family, CF nurse, GP, fertility clinic, pediatrician or books. 71% felt the CF clinician or CF nurse (24%) should be the informer. 57% knew it was rare for a CF man to father a child, 25% knew it was less common to be a father. 24% had had sperm count. 38% expressed wish to have a sperm count. 68% expressed wish to be a father. 43% patients said they would consider fertility treatment. | Patients are not well informed about fertility, most patients wish a sperm count and many patients wish to consider assisted conception. Genetic counseling, screening of the partner for CF carrier status and consideration of implications of raising a child with CF an shortened life span. |
| Nixon et al. [9] AU | Sources of RSH information for girls and their mothers | 55 female patients and their mothers | Questionnaire | CF doctor important source. 87% of girls and 78% of mothers never discussed any of the 7 issues with their CF doctor. 37% felt comfortable asking. 35% were interested but CF team should raise the topic. 19% too embarrassed. | Girls turn to their mother for information, but mothers may not be sufficiently informed. The majority of mothers want more information specific on RSH and CF. CF team should initiate discussion. |
| Sawyer et al. [10] AU | Sexual and reproductive health in men with CF | 94 male patients including 18 transplanted men | Questionnaire | 94% knew about infertility, most hear later (16.4 years) than desired (14.4 years). 53% from preferred source. | Men desired more information on RSH. Earlier discussion is indicated. Semen analysis should be routine. |

(continued on next page)

Table 1 (continued)

| Authors | Topic | Sample | Method | Results | Main conclusions |
|------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------|----------------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| | | | | 53% had semen analysis. 68% of those who did not have semen analysis, wanted this. 73% had semen analysis before the age of 18. In adolescence 1 in 3 assumed not to need condoms. 1 in 10 thought infertility was equal to impotence. 66% wanted more information on reproductive options. 84% wanted children. | Discussion of reproductive options is needed. |
| Frayman et al. [11] AU | RSH in boys with CF: what do parents know and say? | 84 mothers , 64 fathers | Questionnaire | All but one knew about infertility since diagnosis. 19% report subsequent discussions with physician, initiated by parents. Adolescence was considered best time to inform patient. 6% worry about child's reaction. 95% think parents (alone or with CF specialist) should inform son. 30% were satisfied with current knowledge. | Parents are well informed about infertility. Little opportunity but definite need for subsequent discussions about RSH with health care professionals. |
| Gatiss et al. [12] UK | Contraceptive services and advice for women with CF | 42 female patients | Questionnaire | 79% were sexual active. 31% had 19 pregnancies, 5 were unplanned. 50% used contraceptives. 62% reported not to having received contraceptive advice specific to CF. 57% said not to have been warned about antibiotics and combined pill. | Relatively high rate of unplanned pregnancies. Sub-optimal advice or use of range of contraceptive methods. CF team lack in training. Contraceptive services lack knowledge of CF. |
| Korzeniewska et al. [13] POL | Knowledge and experiences of reproductive and sexual health issues in women with CF as well as knowledge and reproductive health attitudes of their parents | 64 female patients and 64 parents | Questionnaire | 68% had first sexual intercourse at a mean age of 19.2 years. 84% of sexual active women used contraceptives. 32.8% understood problems with RSH related to CF for women and men. Sources of information were popular scientific papers and other patients. 70% never discussed RSH with parents. 23% of parents understood female RSH problems related to CF; 56% knew of male infertility; 75% of patients and 40% of parents felt sex health discussions should start between 12 and 14 years of age with CF doctor and mother. | There is a knowledge gap about CF and RSH issues in both female patients and their parents. There is some knowledge about sexual issues, however not enough to have a safe sexual life. |
| Popli et al. [14] UK | Awareness of fertility treatment in men with CF | 37 male patients | Postal questionnaire | 24 knew about infertility problems related to CF. 15 men were first told by their CF physician, 10 knew from a book, CF newsletter of internet, other sources were CF nurse, fertility specialist or parents. 22 were informed during early or late adolescence; 8 first hear when they were in their 20 s and these men considered this too late. Only 50% knew infertility treatment was available. 24 worried about their offspring having CF. 20 worried how CF would affect them being a father. 8 felt is justifiable if they were refused treatment because of their CF. | A significant number of patients do not know about infertility until late in their teens. Many are unaware of treatment options. Worry about CF affecting them being a parent. Worry about their children having CF. Need for proactive reproductive counseling, with access to fertility specialists. |
| Sawyer et al. [15] AU | Variability of male RSH knowledge and preferences and clinical practice across different CF clinics | 5 CF clinics, 264 male patients, including 42 transplanted patients | Questionnaire | 65% knew of near universal infertility, 42% heard about infertility from their preferred source, but later than preferred. Less than half had undergone semen analysis, often later than preferred. 57 men were fathers (29 with assisted reproductive technology). | Men's preferences around RSH were more consistent than clinical practices. Guidelines and training would help reduce the gap between men's RSH preferences and clinical practices. |

Table 2

Important topics in reproductive and sexual health in CF.

| |
|----------------------------------------------------------------|
| Sexual development, puberty delay [2,3,16–20] |
| Semen analysis, small volume ejaculation [2,15,20] |
| Infertility [2,3,14,15,19–22] |
| Menstrual cycle [3,8,17,19,20,23] |
| Condom use—prevention of STDs—birth control [3,12,14,15,23–25] |
| Reproductive treatment options [2,3,14,16,20–22,24–28] |
| Antibiotic use and vulvovaginal candidiasis [14,25,26,29] |
| Pregnancy and CF [3,14,22–24,30–32] |

Including review articles [4,16,24,28,30–32] and guidelines [23].

Information to parents should be provided at diagnosis, preferable early on, but at least during the first 2 years [33]. For parents of boys this generally happens (87%), but no data is available for parents of girls. The majority of parents express a need for re-education when their son is around the age of 10 to be sure they are able to educate their son themselves [9,11].

5. Sources of information

RSH information for girls comes predominantly from their parents and their GP [9,13]. The majority of girls from the Australian study [9] wished to learn more about RSH issues as long as a CF team member raised the topic. However, for modern contraceptives they felt that the CF health care professionals lacked knowledge and preferred to go to their GP, even though their GP may not be up to date with CF care.

Boys generally received information from their parents [5,7,11] and/or a member of the CF team [5,8,10,14]. Those under 16 years were informed more often by their parents, and those over 16 years were informed more often by a health care professional [15]. Other sources of information were CF friends, CF newsletters/websites, a family planning clinic, friends or written material [8,9,13,14]. Fair et al. [6] found that more than half of their patients specifically wanted written information about fertility issues, while less than a quarter remembered receiving any. Both patients and parents expressed the need for multiple sources of information, including written information [9–11,13,15]. Special educational seminars on RSH and CF were requested by the Polish patients and their mothers [13]. However, health care professionals generally reported insufficient training and were uncertain about their skills to talk about RSH in CF [7,11].

6. Attitudes and emotional reactions in communicating about RSH

Embarrassment is a common reaction when discussing RSH issues. Nineteen percent of Australian girls were too uncomfortable to discuss these issues with anyone in the clinic [9]. Seventy percent of Polish girls [13] had never discussed RSH issues with their parents, even though they designated their parents as the preferred source of information. For boys, learning about possible (or reduced) infertility raised strong emotions in 67 of 82 British patients [6]: 40% were shocked,

24% bewildered and 19 were angry. Two Australian studies [10,15] reported that the majority of boys did not remember any strong reaction when told about their infertility, although 32% were a little upset and 18% were very upset. It is important to note that the older the patient the more upsetting the news. The impact of infertility became greater over time especially for men who wanted children. Twenty-two percent of men reported a significant impact of infertility on their relationship.

For parents, Frayman et al. [11] found that 67% of 148 parents reacted negatively to the news about the infertility of their son, but the significance of the news diminished over time. Sixty percent of parents (predominantly mothers) were concerned about their sons' reaction to the knowledge of infertility. Some parents considered it in their child's best interest to inform them early so that there were no surprises when they reached adulthood.

Health care professionals are to some extent concerned about parents' attitudes toward communicating RSH issues [7,11]. Nevertheless, the majority of health care professionals have positive attitudes towards informing patients and parents and see it as their task as CF specialists. Some, be it few, feel apprehensive and consider other health topics more important to talk about and/or believe that medicine is aimed at disease control and less emphasis is needed on other aspects [7,14]. With recent technical developments, e.g., assisted reproductive techniques, professionals are more comfortable to talk about RSH in CF, because they can focus on these techniques [7].

7. Chronic illness and sexual behavior

Laumann et al. [34] argue that sex is a basic human right and a fundamental part of a full and healthy life. For most healthy men and women sex contributes to their personal and relational quality of life. For patients with a chronic disease, sex is also a significant determinant of quality of life, with sexual intimacy being an important mode of communication with their partner [35].

The reviewed papers on RSH tend to focus on fertility and reproductive issues and less so on sexual behavior and/or sexual functioning. Teenagers with CF are sexual active, have a normal sexual libido (despite the delayed onset of puberty) and engage in similar risk taking sexual behavior as their healthy peers (sexual transmitted diseases and unplanned pregnancy) [2]. Apart from anecdotal evidence little is known about the impact of CF on sexual functioning or sexual well-being. From clinical practice we know that some patients describe sex as an efficient and effective method of airway clearance. Female patients have reported that they participate in sexual activity even though they themselves were too tired because of their CF. An exemplar paper on chronic disease and sexual behavior was recently published in the *Journal of Sex Research* [36]. In this paper a conceptual framework was provided to study sexual behavior in relation to chronic illness. The authors discuss 'sexual functioning' (generally accepted as "normal" performance standards described within the sexual response cycle) and sexual well-being (referring to the person's subjective, individual experience of sexuality and how it is evaluated in the context of his or her personal life and relational situation). A chronic disease may affect physical and/or

psychological well-being and therefore indirectly affect sexual behavior. This model is comprehensive and provides an excellent framework for CF research on how CF affects sexual functioning and sexual well-being. As Popli et al. [14] concludes, health care professionals are not only responsible for the prolonging of life, but also to help patients obtain a good quality of life.

8. Methodological and cultural considerations

All but two studies included patients from a single CF clinic (Table 1). No information was given concerning the representativeness of the samples, either nationally or in relation to the general clinic population. Self-report questionnaires were predominantly used for data collection. These questionnaires were ‘ad hoc’ and developed for the purpose of the studies. The different methodologies and populations make it impossible to assess the validity and reliability of the scales used or to reliably compare the results across studies and cultures. Nonetheless, these data have provided a starting point for the discussion of RSH issues and future research.

As the studies originated from the United Kingdom, Australia and Poland, it is important to consider the expected impact of cultural differences in RSH information. General sex education is often first provided in schools [37] and children with CF will receive the same information as their healthy peers. In some countries this education may start in primary school, in other countries this may be in secondary school. The content of this general sexual health education may also differ depending on the cultural and/or religious background of a country. For example, Polish girls with CF felt generally ill informed [13], which may be due to the fact that Poland is a catholic country with mainly traditional catholic education and restrictions on general RSH education in schools.

9. Ethical considerations

Ethical considerations were sparsely mentioned in the studies. An exception is the finding that 8 out of 37 male patients would find it acceptable if they were refused assisted reproductive techniques on the basis of their medical condition [14]. Ethical considerations were discussed by Sawyer et al. [10], for example, issues such as ‘having a child with CF as a parent with CF,’ ‘the process of parents with CF and their partners in balancing the medical needs and shortened life span with the desire to parent children,’ ‘the ethics of partner screening’ and ‘the role of selective termination.’ In their paper on fertility issues in CF, Lyon and Bilton [38] describe the apparent paradox of promoting population screening to reduce the burden of CF disease while supporting men with CF to parent children who will be carriers. It is beyond the scope of this review to discuss ethical issues in detail. A recent article discussing the ethics of patient and parental procreative obligations in CF is suggested for further reading [39]. One has to be aware of the impact ethical considerations may have on the availability of information. For example, the personal views of health care professional may result in some patients not receiving the information they need.

10. Conclusions

We reviewed 11 research papers on education and communication about reproductive health and sexuality in CF published over the past 10 years. The studies have used different methodologies and populations and have originated from different cultural backgrounds making them difficult to compare. More studies have been undertaken with male patients than female patients. This may be because of the immediate impact of infertility on men with CF. Nevertheless, the studies including females revealed that women have a similar need for information, and that the information that they have received often lacks a specific CF focus.

There is wide variability concerning the timing of information given to patients with CF. All reviewed studies have indicated that health care professionals in CF clinics inform later than is preferred by the patients, which would be around 13 years of age. Attitudes, emotional barriers, embarrassment and personal preferences seem to play an important role in communication about RSH in CF [6,9–12,15], which may lead to under-information and negative emotional reactions.

Several recommendations can be drawn:

- Women need more detailed CF-related information, for example about side effects of CF drugs or the raised risk of thrombosis when taking oral contraceptives in combination of having an indwelling intravenous catheter.
- Men need present-day information which should include new topics, for example assisted reproductive techniques.
- Possible misconceptions need permanent attention, e.g., men mixing up infertility and impotence.
- Health care professionals should initiate discussions with patients and their parents.
- Provide information during early during adolescence.
- Repeat information over time, perhaps at set stages within child/adolescents development.
- Need for multiple source of information, including up-to-date written information and access to fertility advice centers.
- Parents need help to fulfil their role to inform and educate their children.
- When informing patients about RSH issues in CF, one has to take into account sexual experiences and sexual well-being, with respect for patients’ privacy.
- Ethical and cultural issues need to be considered when informing patients with CF and their parents about reproductive and sexual health issues.

10.1. The need for guidelines

Looking at the studies over the past 10 years and comparing the data with earlier studies we have to conclude that there are areas where patients are still ill informed about RSH in relation to their CF [12–15,40,41]. Patients with CF understand many aspects of their disease [42], but within the area of RSH there are important knowledge gaps. Themes of communicating and informing about RSH in CF are broadening, but the issues of ‘timing,’ ‘who informs,’ ‘who follows up’ and ‘what is communicated or not’

remain important. Consensus papers have been written on the treatment of CF [43,44] and CF pregnancy [23], but no guidelines are available on how to counsel sexually active males and females with CF. Cultural differences will impact on the extent to which guidelines on RSH in CF will be implemented in different countries and/or amongst different cultural groups.

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