Sexual Health in Cystic Fibrosis

Produced By:

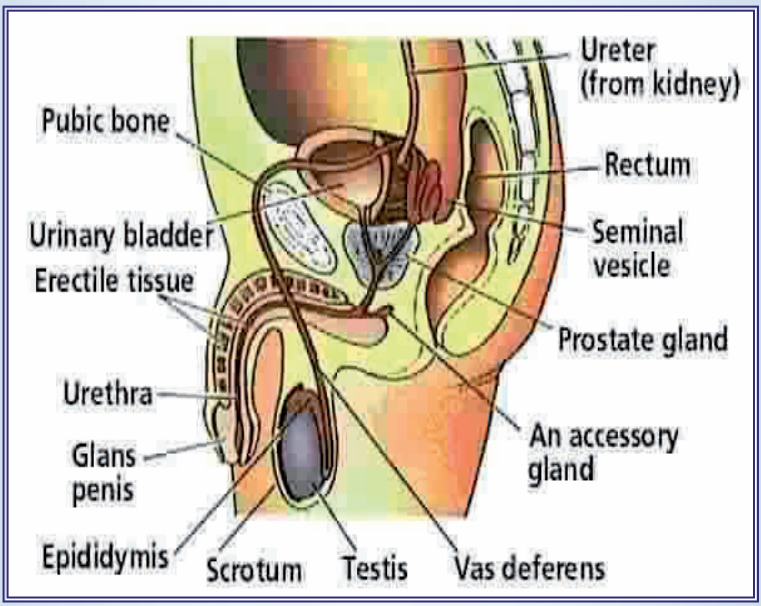
Amandeep Kaur Mann, Sanjay Haresh Chotirmall,
Cedric Gunaratnam and Noel Gerard McElvaney

Cystic Fibrosis & Sexual Well Being

Cystic Fibrosis (CF) is an inherited multi-organ disease that is caused by an abnormality of the CF gene. This causes your body secretions to be more thick than normal, leading to, for example, chronic lung infections (thicker mucus). CF may also cause problems with your intestine, pancreas, bones, or liver.

Whilst many people with CF enjoy normal sexual development, a delay in puberty onset may be experienced. However, sexual activity and libido are no different than those of the same age. Although prospects of parenthood are not impossible, having CF may hinder your ability to have children.

Ongoing research, and a greater understanding of CF, are allowing physicians to find newer methods to help you have a smoother reproductive life.

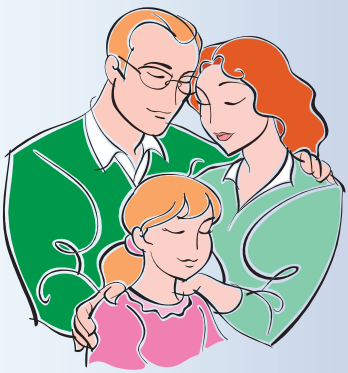


The Male Reproductive System

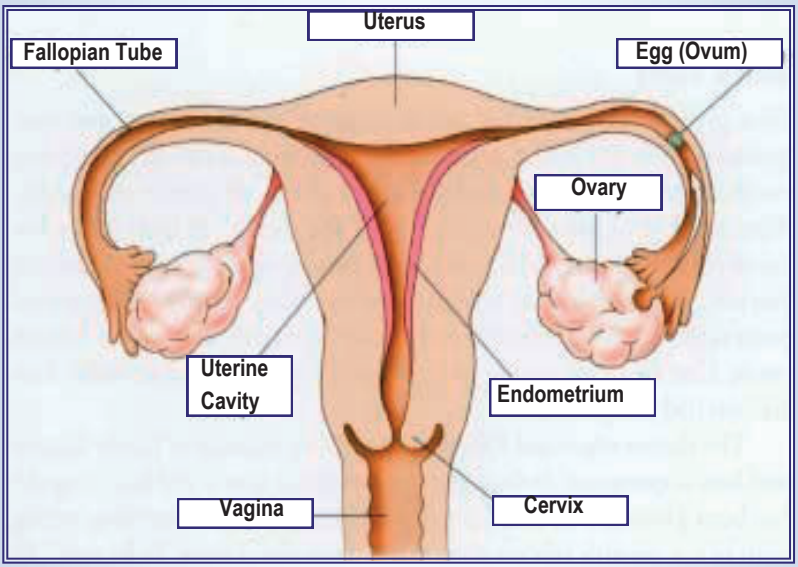
Male Fertility in Cystic Fibrosis

Regardless of CF disease severity, 96-98% of males are infertile[1]. This is usually due to an absence of a tube called the *vas deferens.* The *vas deferens* delivers sperm from the testes to the penis, and eventually into the female partner. Therefore, it can be said that male infertility is due to a failure of sperm delivery, and not an abnormality of the sperm themselves. Some medical advances have been made in order to help men with CF father children.

For instance, a process known as *in-vitro* fertilization (IVF) can assist men with CF in becoming fathers. In this process a fine needle is inserted into the epididymis (*microsurgical epididymal sperm aspiration; MESA*) and sperm are retrieved. The sperm are mated with eggs taken from the female partner, (*intracytoplasmic sperm injection; ICSI*) and when ready, the embryo is inserted into the female. Men with CF who underwent MESA, along with one or two cycles of ICSI, in one study, had a success rate of 63%[1].



Whilst prospects of fatherhood for men with CF are currently good, it is important to discuss specific health concerns with your CF medical team.



The Female Reproductive System

Pregnancy & Cystic Fibrosis

Women with CF generally have a healthy reproductive cycle. They may, however, experience difficulty in becoming pregnant. Unlike men with CF, there are no abnormalities in the delivery mechanism of the egg within the female reproductive system.

However, the cervical fluid of women with CF is thicker than usual. This increased thickness makes the journey of the male sperm to the female egg within the uterus much more difficult[2]. The thick bodily fluids within the cervix may also make ideal breeding places for various bacteria. Therefore, it is essential to maintain a high standard of self-hygiene as well as follow any advice offered to you by your healthcare professional.

Some women with CF who become pregnant may have healthier lives and a better survival rate at 10 years than those women who are not pregnant[3]. However, it is important that CF women plan their pregnancies with the support of their CF multidisciplinary care team, and involve an obstetrician as early as possible, as CF pregnancies may require special monitoring and care[4].

Cystic Fibrosis & Sexual Safety

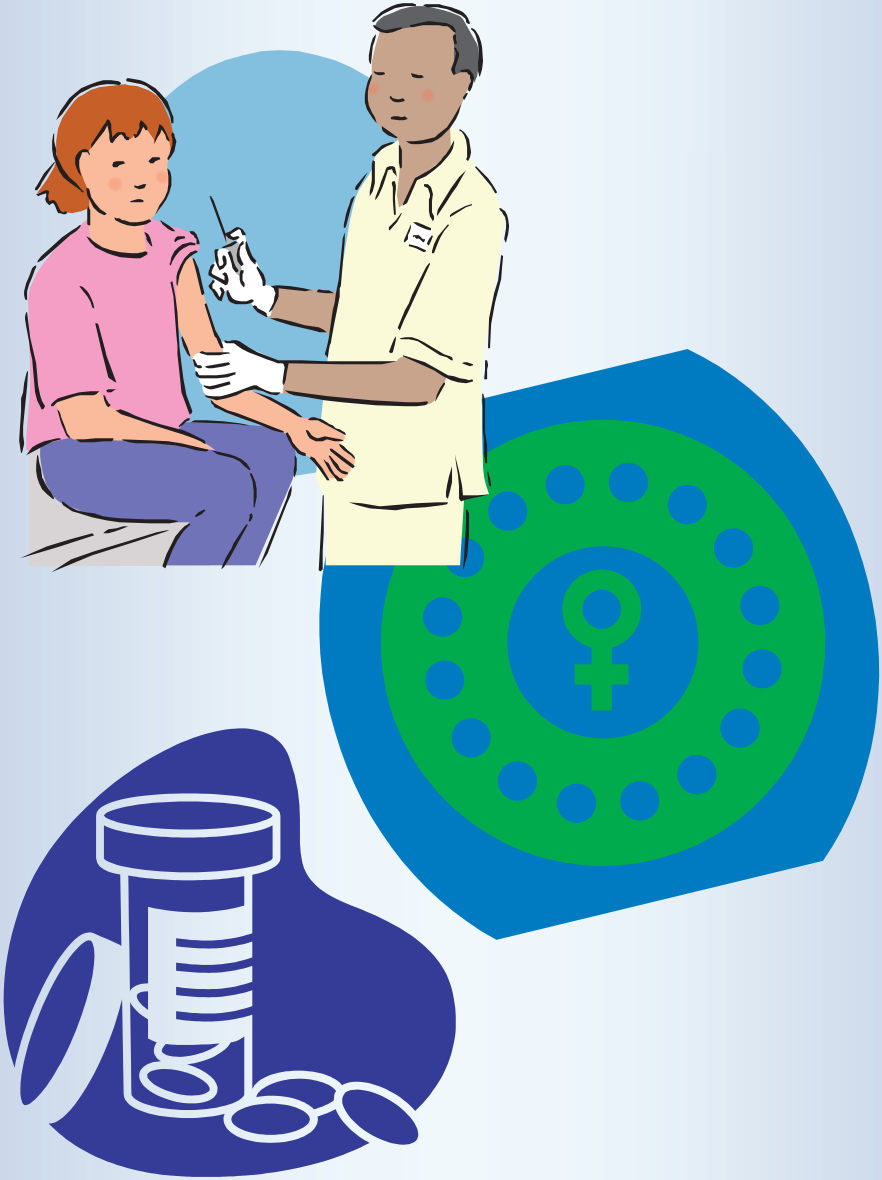
Although it would be difficult for many men with CF to become biological fathers without medical assistance, and while females with CF may have difficulty in conceiving, practicing safe sex is still vital for your well-being.

Sexual safety is important in preventing the spread of sexually transmitted diseases (STDs). STDs include:

- Chlamydia
- Genital warts
- Gonorrhea
- HIV and AIDS
- Syphilis
- Hepatitis B and C

STDs, if left untreated, can cause many complications in both women and men, including infertility and cancer of the reproductive tracts. The best way to combat STDs is through prevention. There are many ways to prevent STDs, for example:

1. Commit to sexual relationship with a single person
2. Use a good quality condom (warning: it is not 100% protective)
3. Don’t use an oil based lubricant when using a condom
4. Keep good hygiene



Cystic Fibrosis & Contraception

Currently, many methods of contraception are available. These include:

- Male and female condoms
- Oral contraceptive pill (OCP)
- Intra-uterine devices (IUD)
- Vaginal rings
- Hormones delivered via a patch (Evra)
- Progestogen injection or implantation of a device secreting it slowly (Mirena Coil)

Despite the OCP being a popular contraception method amongst adolescent females, concerns regarding its efficacy perhaps due to interactions with other medications and reduced absorption in CF females evoked some hesitation in its use. However, recent literature suggests that there is no major overall difference between its efficacy in CF females, and non-CF females[5].

Females with CF are advised to speak to their physicians about appropriate contraceptive methods, as certain methods may prove much more beneficial than others.

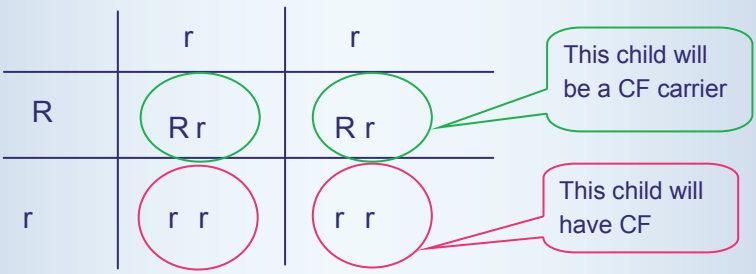
Cystic Fibrosis & Genetics

Currently, in the Republic of Ireland 1 in 1,461 newborns are born with CF each year[6]. CF is one of the most common genetically inherited life threatening diseases in Ireland. It is estimated that every 1 in 19 people in Ireland are carriers of a defective CF gene[7]. A gene can be thought of as a basic building block of what a person looks like (i.e. hair colour), or how their body functions. Over 700 alterations to the normal CF gene have been identified to date.

Diseases can be inherited in many ways. Two common ways are autosomal dominant inheritance, and autosomal recessive inheritance. CF is a disease that is inherited in an autosomal recessive pattern. Everybody inherits two copies of the CF gene, cystic fibrosis transmembrane regulator (CFTR). Those with cystic fibrosis inherit two *altered* copies of the CF gene. A CF carrier is someone who inherits only one defective copy of the CF gene. In autosomal recessive inheritance, in order to have the disease you must inherit two altered copies of the gene, therefore, a carrier will not show signs of CF as they only have one altered copy of the gene.

Parent with CF gene carrier

Parent with CF



In the example above, the person who has CF is represented by rr, and the CF carrier, who doesn't have CF, but carries one copy of the altered CF gene, is represented with Rr.

As you can see, the chances of having a child with CF are 50% (there are 2 rr out of the 4 possibilities), and there is a 50% chance that the children will be carriers of an altered CF gene.

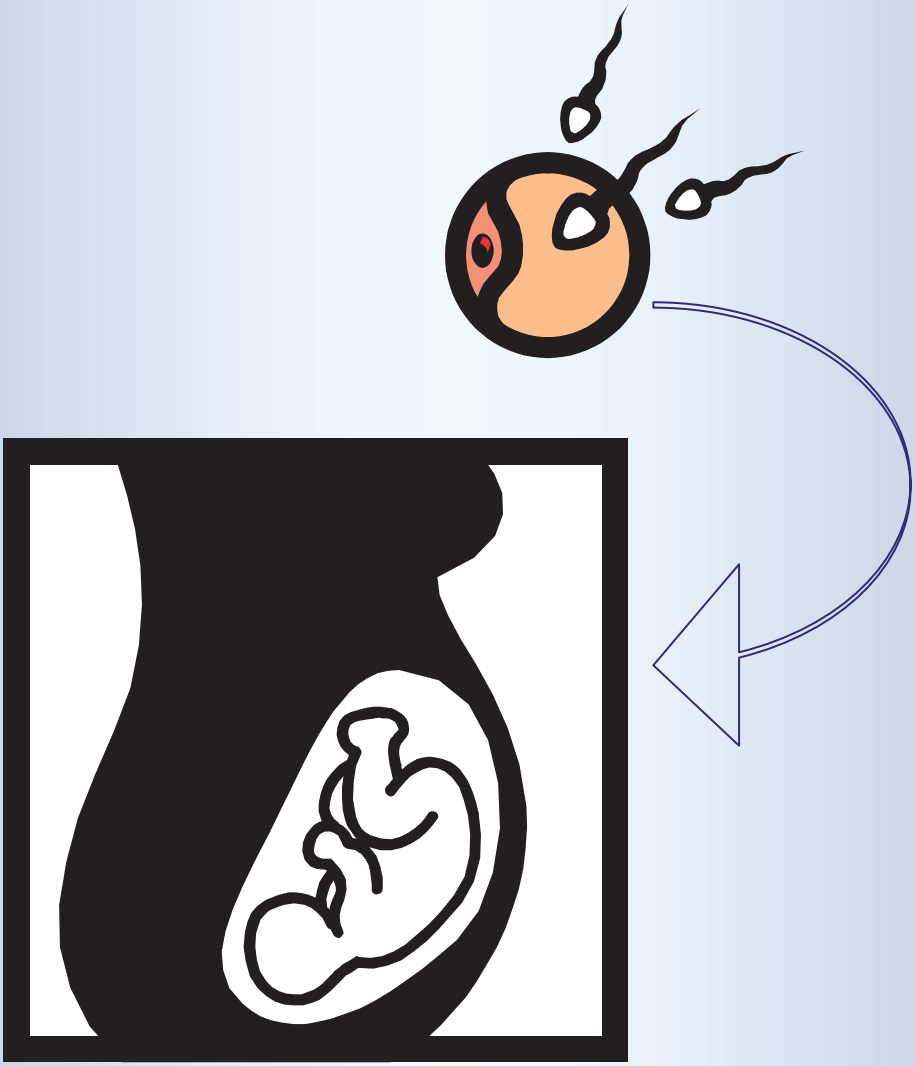
Will My Child Have Cystic Fibrosis?

The probability of having a child with CF is determined by the genetic status of the parents. Probabilities change greatly depending on if one or both parents are carriers, or is a person with cystic fibrosis (PWCF). Below is an example for some of these situations.

1. Both parents are carriers of an altered CF gene:
    - 50% chance the child being a CF carrier
    - 25% chance the child will have CF
    - 25% chance the child will not have CF nor be a carrier

2. One parent has CF, and another parent is a carrier:
    - 50% chance the child will have CF
    - 50% chance the child will be a CF carrier

3. Both parents have CF:
    - 100% chance the child will have CF



Cystic Fibrosis & Genetic Counselling

People living with CF who want to have children can approach their healthcare team, who, in turn, will refer them to the appropriate services.

Depending on your needs as a CF patient, certain services may be undertaken. There are specialized units that deal with genetics and fertility issues. One such unit exists in Rotunda Hospital in Co. Dublin. The *Human Assisted Reproduction Ireland* (HARI) unit in Rotunda was opened in 1989, and since then has been serving many couples who have had problems with fertility. The HARI unit offers a wide variety of services including:

- Intrauterine Insemination (IUI)
- In Vitro Fertilisation (IVF)
- Testicular Sperm Extraction (TESE)
- Intra-Cytoplasmic Sperm Injection (ICSI)

For further information, please visit the HARI unit website, www.hari.ie .

The *National Centre for Medical Genetics* seeks to provide a comprehensive service for all patients and families in the Republic of Ireland affected by or at risk of a genetic disorder. The Centre is based at Our Lady's Children's Hospital Crumlin, Dublin. The Centre provides a service for both children and adults. The Centre provides a clinical service for individuals and their families who are affected by, or at risk of a disorder with a significant genetic component such as cystic fibrosis.

For further information visit www.genetics.ie

References:

1. McCallum TJ, Milunsky JM, Cunningham DL, Harris DH, Maher TA, Oates RD. Fertility in Men With Cystic Fibrosis: An Update on Current Surgical Practices and Outcomes. Chest 2000; 118:1059-62.

2. Lyon A, Bilton D. Fertility Issues in Cystic Fibrosis. Paediatr Respir Rev 2002; 3: 263-240.

3. Goss CH, Rubenfeld GD, Otto K, Aitken ML. The Effect of Pregnancy on Survival in Women with Cystic Fibrosis. Chest 2003; 124: 1460-68.

4. Edenborough FP, Borgo G, Knoop C, Lannefors L, Mackenzie WE, Madge S, Morton AM, Oxley HC, Touw DJ, Benham M, Johannesson M. Guidelines for the Management of Pregnancy in Women with Cystic Fibrosis. J Cyst Fibros 2008; 7 (1): S1 –S32.

5. Plant BJ, Goss CH, Tonelli MR, McDonanld G, Black RA, Aitken ML. Contraceptive practices in Women with Cystic Fibrosis. J Cyst Fibros 2008; 7 (5): 412-414.

6. Cystic Fibrosis Registry of Ireland. Annual Report 2006. Available from: http://www.cfireland.ie/publications/annual_reports/annual_report_2006_registry.pdf

7. Cystic Fibrosis Association of Ireland., Dublin; 2008 [updated 2008; cited 2009 08/03/2009] Available from: http://www.cfireland.ie/articles.php/what_is_cf?

Notes

Questions

Please feel free to contact:

Acknowledgements:

This work would not have been possible without the help from our CF multi-disciplinary team at Beaumont Hospital, Dublin, Ireland. We appreciate and acknowledge our CF nurse specialists for all their input (Cassandra O'Donohoe, Anne Marie Lyons, and Claire Bolton). We are indebted to our CF patient cohort who provided invaluable feedback during the conception and revision of this publication. Thank-you to the Cystic Fibrosis Association of Ireland (CFAI) for the support in printing and distribution of this booklet.