

#### Inside This Issue

- 1-2** CF & Fertility
- 2** Great Stride Walk
- 2** H&L CF Team
- 2** Important Dates  
May is CF Awareness  
Month!

## Cystic Fibrosis and Fertility

Cystic fibrosis (CF) is caused by mutations in the gene responsible for producing the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) protein. The CFTR protein helps to maintain the balance of salt and water on many surfaces of the body, such as the surface of the lung and pancreas. When the protein is not working correctly, chloride- a component of salt- becomes trapped in cells. The trapped chloride prohibits cell surfaces from becoming hydrated as water cannot flow properly through the cells. This causes the mucus covering the cells to become thick and sticky.

In people with CF, the thickened mucus can cause issues with many of the body's processes. This includes fertility, or the ability to produce offspring through reproduction. The good news is, with advancements in treatments and CF care, many people with CF can have biological children. As people with CF live longer, healthier lives than before, more people will need to make decisions about their reproductive health. The following is some information on how CF affects fertility in men and women, as well as information on some of the advancements in treatment that have been made to help improve fertility.

#### CFTR Mutation Effects on Fertility in Women:

- Thicker cervical mucus – this makes it difficult for sperm to cross the cervix to fertilize an egg.
- Irregular ovulation – most commonly due to poor nutrition. A low quality diet can affect the levels of the hormones responsible for ovulation (or when the uterus releases an egg).
- It may take longer to conceive
- More prone to vaginal candida – this is due to antibiotic and steroid use as it can change the levels of acidity and levels of bacteria.
- Stress incontinence – frequent coughing can weaken the pelvic floor muscles or the muscles used in childbirth.

Most women can become pregnant!

#### CFTR Mutation Effects on Fertility in Men:

- 97-98% of men are infertile but not sterile- meaning they are still able to produce sperm.
- Men with CF are infertile due to absence of the sperm canal (also known as congenital bilateral absence of the vas deferens or CBAVD).
- The cause of CBAVD is unknown, however it is thought to be related to mutations in the CFTR gene.
- An absence of the sperm canal leads to sperm not being able to make it to the semen.
- Sperm production is normal in 90% of men with CF.
- Contraception is recommended unless infertility is confirmed.
- Puberty may be delayed in adolescents– often caused by impaired production of hormones. Hormone production can be affected by poor nutritional status.

Most men with CF will have normal testosterone production, and will achieve normal height, muscle mass, and other sexual features.

## Cystic Fibrosis and Fertility Continued

### Effects of CFTR Modulators of Fertility:

The full effects of CFTR modulators on fertility has not yet been fully studied (it is in the process)

- However there seems to be growing cases of women becoming pregnant after starting a CFTR modulator, which may suggest improved chances of pregnancy in women with CF
  - This is due to CFTR modulators being thought to thin the cervical mucus and normalize the pH of cervical mucus
  - CFTR modulators may help improve the general health and nutritional status of those with CF
- Up till now –no observed negative effects on babies conceived or during pregnancy when mothers stayed on modulator therapy
- Further studies are ongoing:
  - PROMISE study – focusing on how Trikafta affects overall health in people with CF
  - Slight increase in pregnancy rate shown with introduction of Kalydeco and Symdeco
  - Expecting to see a Trikafta Boom this year!
- In men, CFTR modulators may help increase ejaculate volume – still does not change the vas deferens
- 25-50% of pregnancies in women are unplanned
  - Birth Control must be used!!
  - Only Orkambi has been found to interact with hormonal contraception and those taking Orkambi should use another form

### Options, Choices and Treatments

- Assisted Reproductive Therapy (ART)
- In-vitro fertilization
  - Sperm retrieval/aspiration with subsequent in-vitro fertilization with embryo.
  - Success rates are 20-40%
  - Costly, physically and emotionally challenging
- Intrauterine insemination (IUI) – sperm is placed inside a woman’s uterus (AKA artificial insemination)
  - increases the number of sperm that reach the fallopian tubes
- Use of donor sperm, eggs and embryos
- Gestational surrogate – egg and sperm are from the intended parents and create an embryo for implantation
- Adoption

## Recent Event: Great Strides

Great Strides is the Cystic Fibrosis Foundation’s largest national fundraiser. The event is held every year to help raise awareness and fund research for a cure for CF. Great Strides provides a fantastic opportunity for family, friends, students, and colleagues to come together and make a difference. This year, the local Great Strides event was held in Moorhead, MN at the M.B. Johnson Large Shelter on April 30<sup>th</sup>, 2022. If you are interested in walking next year or wish to donate to a person or team, please visit

[https://fightcf.cff.org/site/TR/GreatStrides/80\\_Minnesota\\_Minneapolis?fr\\_id=9106&pg=entry](https://fightcf.cff.org/site/TR/GreatStrides/80_Minnesota_Minneapolis?fr_id=9106&pg=entry) for more information.

## Important Dates:

Cystic Fibrosis Clinic Dates  
 May 19, 2022  
 June 16, 2022  
 July 21, 2022  
 August 18<sup>th</sup>, 2022

Patient Advocacy Council Dates TBA  
 Check your e-mail

Your CF Team Members:  
 Dr. Monica Paulo  
 Dr. Jane Gorud  
 Dr. Parveen Wahab  
 Deb Fueller FNPC  
 Heidi Reiter FNPC  
 Leanne Saville PharmD  
 Stephanie Friedt RRT.RPFT  
 Amber Burck, LPN  
 Samantha Snow, RDN, LRD  
 Christina Bond, LMSW

36<sup>th</sup> North American Cystic Fibrosis Conference  
 November 3<sup>rd</sup>- November 5<sup>th</sup>, 2022 in Philadelphia, PA  
 More information is available at <https://www.nacconference.org/Home/>