



PUBERTY AND FERTILITY IN CF

Puberty* is the period of time when a person's body changes and becomes sexually mature. Puberty occurs when certain **hormones*** (special chemicals in the body), such as **estrogen***, **testosterone***, and **growth hormone***, are made by the body. This "surge" or increase of hormones sends signals to the body and leads to dramatic physical and emotional changes.

Adolescence* is the stage of development when puberty takes place. At no other time in a person's life is there such intense growth and development, except during infancy. Although we talk about "reaching puberty," it doesn't happen overnight—puberty is actually a process that takes three to five years. No two **adolescents*** go through puberty exactly the same way nor do they reach puberty at the same age. Although changes in the body are most noticeable, an adolescent's brain is maturing during puberty as well, and will continue to mature after the body is fully developed. With these brain and body changes in puberty, adolescents' interests change and most become more focused on dating, relationships, appearance, and sexuality.

A person normally becomes **fertile*** during puberty. **Fertility*** is a woman's ability to **conceive*** (become pregnant) or a man's ability to bring about **conception*** (getting someone pregnant). Sometimes certain conditions or diseases can affect the onset of puberty. **Cystic fibrosis*** (CF*) can affect the timing of puberty and fertility in both males and females.

GROWTH AND DEVELOPMENT IN PUBERTY

Young people with CF go through puberty and develop into men and women just as other people without CF do. Adolescents with CF, however, may start puberty later than other adolescents. Some of the physical signs of growth and development that may be delayed include:

- Getting taller or reaching close to adult height
- Underarm hair, pubic hair, and facial hair (in boys)
- Boys' voices getting deeper and their **genitals*** growing larger
- Girls developing breasts and beginning to have **menstrual periods*** or **menses***

When puberty starts may depend on the overall health of a person. A person with CF who is underweight, for example, or who has severe lung disease may go through puberty later than other adolescents with CF. Severely underweight girls may begin their periods even later, or their periods may not follow a regular cycle. Having enough body fat helps puberty start on time. Adolescents with CF who have good nutrition will usually have a normal growth spurt. However, some children with CF, even when well-nourished, tend to start puberty late. A reason for this may be how a person with CF makes and uses growth hormones. If your child does not show any signs of puberty by age 14 for boys (an increase in size of the **testes*** is the first sign) and age 13 for girls (breast development is the first sign), talk with your doctor. Your child may need to see a pediatric **endocrinologist***, a doctor who specializes in hormone problems.

Your son or daughter may find it hard to start puberty later than friends and classmates. Your child may be self-conscious about being one of the shortest kids in his or her class or being less developed than the other kids in the locker room. If your child is struggling with feelings about puberty or how he or she looks, talking with a CF health care team member may help your child figure out ways to improve nutrition or general health to speed up the start of puberty. The CF health care team can also help reassure your child that the time will come for him or her to go through puberty—it will happen.

MALE FERTILITY

During puberty, boys with CF develop normal physical sexual features. They produce sex hormones and have normal sexual drives. More than 90 percent of men with CF, however, are **infertile*** and cannot make a female pregnant. Being infertile does not mean that a man is impotent (unable to have **sexual intercourse***). Men with CF can have sexual intercourse and produce **sperm*** like other young men. But in CF the tube that carries the sperm from the testes to the **seminal vesicles*** (called the **vas deferens***) is blocked or absent. When the vas deferens is absent, it is called **congenital bilateral absence of the vas deferens*** (CBAVD). If a man has CBAVD, his **semen*** (the fluid **ejaculated*** from the **penis*** when sexually stimulated) contains no sperm. For pregnancy to occur, a man's sperm **cell*** must meet a

*See CF Words to Know Glossary.



woman's egg cell. To know whether a man is infertile, the doctor can do a "sperm count" on semen ejaculated from the penis. A man with CF can talk with his doctor about having this test. Up to 5 percent of men with CF do have sperm in their semen and can father children naturally.

New medical techniques allow some men with CBAVD to father children. There is now a way to get the sperm from the testes and **fertilize*** the egg from a female. A fertility **specialist*** can give men with CF more information about how it is done and the success rate.

FEMALE FERTILITY

In puberty, females begin to **menstruate*** (monthly flow of blood). **Menstrual cycles*** usually last 28 to 31 days, but they can vary from woman to woman and from month to month. During the first half of the cycle, the **uterus*** prepares itself for pregnancy by building up extra tissue. If the woman does not get pregnant during this time, the uterus releases or sheds this extra tissue and some blood. This release is called having a menstrual period or menses. After the menses is over (usually in 4 to 7 days), the menstrual cycle begins again.

Once they have had their first period (menses), many females with CF are able to get pregnant. Some women with CF do have children of their own. Others, however, have problems getting pregnant. Although **ovulation*** (release of eggs from the **ovary***) occurs normally in CF, the **mucus*** in the woman's **cervix*** can be

abnormally thick. Very thick mucus makes it hard for a male's sperm to get through the cervix to meet the egg. Also, women with CF who are very underweight can have less frequent and less regular ovulation, making it more difficult to get pregnant.

PREGNANCY

Getting Ready for Pregnancy

When a woman with CF is considering having a baby, she should talk with her CF doctor about the risks, and look at her current **lung function*** and weight to see if she can have a healthy pregnancy. Ideally, a woman with CF should be at or close to her ideal body weight before getting pregnant. A woman with CF has to take in an extra 300 **calories*** every day and gain about 20 pounds during a pregnancy to ensure normal growth of the baby. Being at one's ideal body weight reduces the risk of having a premature baby or needing a **cesarean section*** (surgery) to deliver the baby.

Certain medications could put the developing baby at risk; therefore, the CF health care team may need to change some of the woman's CF medicines *before* she gets pregnant.

Health Risks with Pregnancy

Pregnancy can be more risky in women with CF who have lung problems. Pregnancy can sometimes lead to more lung problems. When a woman with CF is pregnant, it is important for her to watch for signs of **infection*** and to take action to prevent lung problems with treatments (**airway clearance*** and **breathing treatments***). A woman who has

severe lung disease may not have enough lung function to have a healthy pregnancy. In this case, pregnancy could put both the woman and her baby at risk.

Some women with CF develop **diabetes* (blood sugar* problems)** during pregnancy and need to control it for their health and their babies' health. If a woman with CF is pregnant, she needs to get medical care from an **obstetrician*** (OB) who specializes in high-risk pregnancies. A high-risk obstetrician can help treat diabetes in pregnancy and can help prevent or treat other health problems related to the pregnancy.

Besides seeing the obstetrician, pregnant women with CF should continue to see their CF health care team for regular CF care.

PASSING ON THE CF GENE

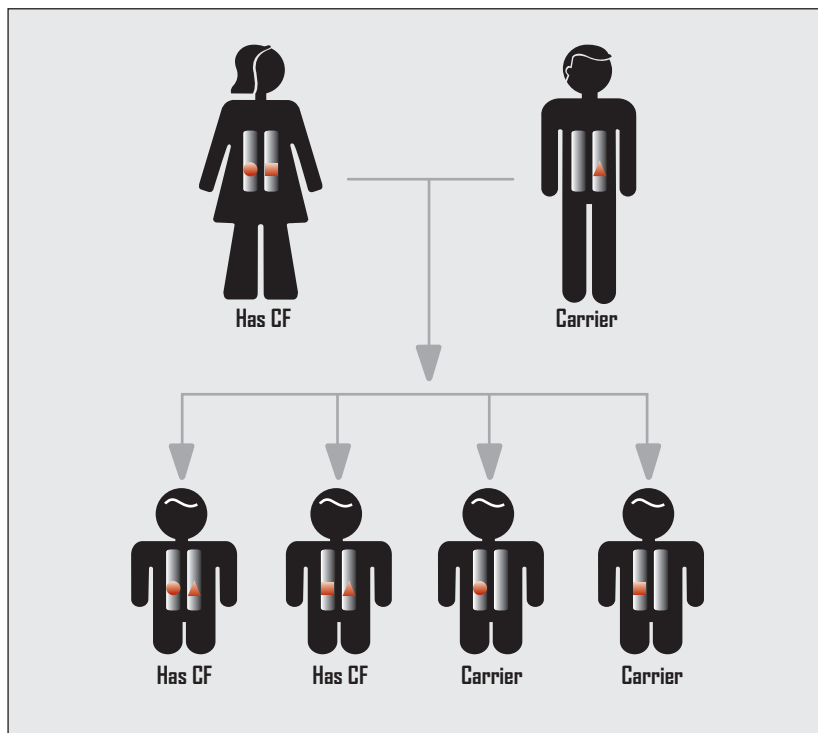
Along with planning a pregnancy like other parents, people with CF have other things to think about, such as: What is the possibility I could pass the CF **gene*** on to my child? And, what are the chances my child will have CF? When a person with CF has a child, the baby will get a CF gene from the parent with CF. The child will either be a **carrier*** of the CF gene or will have CF. Whether the child is a carrier or has CF depends on the other parent. Remember, a person has to **inherit*** two CF genes to have the disease.

Here are the possibilities:

- When the other parent is a CF carrier (has one CF gene), the child has one chance in two (50%) of having CF.



INHERITING CF: PARENT WITH CF AND PARENT CARRYING CF GENE



- When both parents have CF, every child they conceive will have CF.
- When the other parent does not have CF and is not a CF carrier (has no CF genes), the child will be a carrier of the CF gene but will not have CF.

It is very important that a person with CF have his or her partner tested to see if the partner is a carrier for any CF gene. **Prenatal*** testing of the unborn baby (done during pregnancy) can usually tell whether the baby has CF. For more information, see “THE GENETICS OF CF” in the appendix of the CF FEP module *Beginning CF Care* and talk to the CF health care team.

BIRTH CONTROL AND CF

Women with CF who do not want to get pregnant need to either not have sexual intercourse (**abstinence***) or use some form of birth control. Birth control, or

contraception* is used to prevent pregnancy. Several methods of birth control are available:

- Hormone methods, such as pills, a skin patch, or a device that goes in the **vagina*** (vaginal ring) or uterus (intrauterine device or IUD)
- Barrier methods that keep sperm from getting to the egg, such as condoms, a diaphragm (a special cap that is put into the vagina to cover the cervix), or a copper IUD
- Surgery to permanently prevent pregnancy, such as a tubal ligation (the **fallopian tubes*** of a woman are tied so that the eggs cannot pass through the tubes to the sperm)

Before becoming sexually active, a woman with CF should talk with her CF doctor or **gynecologist*** (doctor who specializes in women’s health) about using birth control. She should have a **pelvic**

exam* before starting birth control and talk with her doctor about the risks of each type of birth control method. For example, birth control pills may cause an increase in blood sugar levels if a woman has **CF-related diabetes (CFRD)***. Birth control pills may also have drug interactions with certain **antibiotics*** or other medicines.

Keep in mind that abstinence (not having sexual intercourse) is 100 percent effective for both protecting against **sexually transmitted diseases*** (STDs, sometimes called sexually transmitted infections or STIs) and preventing pregnancy.

SEXUALLY TRANSMITTED DISEASES

Whether or not a man or woman with CF is fertile, he or she can still get and pass on an STD. Sexually transmitted diseases are infections passed from one person to another through sexual activity. Examples of STDs include chlamydia, HPV (human papillomavirus), syphilis, and gonorrhea. Other infections that can be passed from person to person through sexual activity include infectious hepatitis, human immunodeficiency virus or HIV (the virus that causes AIDS), and herpes. Often people cannot tell if their partners have an STD by just looking at them. Many times people are infected without knowing it, but these infections can cause serious health problems.

The best protection from STDs, of course, is to not be sexually active. If a person is sexually active, condoms can help protect against STDs. To be protected, a couple has to use a condom every time they have intercourse (sex). There is no “safe” sex, just “safer” sex.

*See CF Words to Know Glossary.